CARDIAC CATHETERIZATION IN CONGENITAL HEART DISEASE

LONDON
GEOFFREY CUMBERLEGE
OXFORD UNIVERSITY PRESS

CARDIAC CATHETERIZATION IN CONGENITAL HEART DISEASE

A CLINICAL AND PHYSIOLOGICAL STUDY IN INFANTS AND CHILDREN

ANDRE COURNAND, M.D.
Associate Professor, Department of Medicine
College of Physicians and Surgeons
Columbia University

JANET S. BALDWIN, M.D.
Assistant Professor, Department of Pediatrics
New York University College of Medicine

AARON HIMMELSTEIN, M.D.
Instructor, Department of Surgery
College of Physicians and Surgeons
Columbia University

THE COMMONWEALTH FUND · NEW YORK 1949

FIRST PRINTING MARCH 1949
SECOND PRINTING APRIL 1950

PUBLISHED BY THE COMMONWEALTH FUND
41 EAST 57TH STREET, NEW YORK 22, N Y

PRINTED IN THE UNITED STATES OF AMERICA
BY E L. HILDRETH & COMPANY, INC

To the memory of
Pierre Birel Rosset Cournand

*"Having fulfilled his course in a short time,
he fulfilled long years."*

CONTENTS

INTRODUCTION

IN recent years the technique of right heart catheterization has been increasingly applied to the diagnosis of congenital malformations of the heart and large vessels In 1945–46 Brannon, Weens, and Warren reported the use of this technique in the diagnosis of auricular septal defects, and E de F Baldwin, Moore, and Noble reported its use in the diagnosis of interventricular defects Since then *two other groups working under the direction of Dexter and of Bing, have reported* their findings in various types of congenital cardiac malformations and have described in detail the procedure followed in their studies The contribution of both groups has been outstanding and their published papers should be read by anyone interested in this field of investigation *

So far, however, there has been no concerted effort by clinicians and investigators to pool the results of their separate studies, to compare and to integrate clinical and physiological data This monograph is presented as an attempt in such a direction

A study of a varied group of congenital defects of the heart was begun by the present authors in January 1946 as a joint undertaking between the Pediatric Service and the Children's Cardiac *Clinic of Bellevue Hospital (New York University Division)*, the Children's Cardiac Clinic of Lenox Hill Hospital, and the Cardio Pulmonary Laboratory of the Chest Service of Bellevue Hospital (Columbia University Division) One of the participants was especially interested in pediatrics and cardiology, while the others had had experience with the technique of cardiac catheterization in problems related to the physiology of the circulation in adults In the present investigation it was felt particularly important to explore the possibility of extending the method of cardiac catheterization to the study of young children and infants But in order to achieve this it was found necessary to introduce two new features Because of the small size of the arm vein in many cases, the saphenous vein at the femoral region was chosen as the site for introducing the catheter *into the venous system Also,* owing to lack of voluntary cooperation in the younger age group and the necessity of maintaining a steady state throughout the period of study the use of general anesthesia was adopted In view of the need of an anesthetic which would not interfere with gas analysis, avertin was tried and found satisfactory

The essential purpose of a combined study was to make observations of a physiological nature in various types of congenital anomalies of the heart, and to compare and correlate these observations with physical signs and x ray findings It was expected to gain additional diagnostic information and a better understanding of the physiological factors underlying clinical observations as well as to add to the knowledge in the field of the physiology of the circulation

This monograph is divided into two parts In the first part the methods used in the physiological studies are described and discussed In the second part individual cases illustrating various types of malformation are presented

Some of this material was exhibited at the International Congress of Pediatrics held in New York City in July 1947 On this occasion all the data were presented in a simple schematic form and were correlated and compared The method of presentation proved so helpful in demonstrating the respective values of various aspects of the clinical and physiological studies that it has been retained in this monograph several cases having been added to the original material The cases were chosen in order to give a general idea of the most common problems we have encountered in this field of investigation

In view of the fact that extensive studies of the cyanotic group are being carried out by Bing at the Johns Hopkins School of Medicine the emphasis here was deliberately placed on the other types of malformation of the heart Surgical and pathological confirmation of the diagnosis has

* See Bibliography, pp 107–108

been available only in certain of the cases studied. We therefore consider that the presentation we are making at this time, with the above limitations, is in the nature of a pilot endeavor. It is hoped that the results of other investigations carried out along similar lines in a greater number and larger variety of cases, and with surgical or pathological data, will be made available to those interested in this group of fascinating but often baffling cardiac conditions.

We wish to acknowledge our indebtedness to Dr. Hurley L. Motley, who analyzed most of the blood pressure tracings and mounted them for reproduction; to Dr. Lars Werkö, Dr. David T. Dresdale, and Dr. Herbert M. Weiner for their contribution in recording the blood pressure tracings; to Mrs. Marianne S. Lester, Miss Blanche Spierto, and Miss Dorothy Allen, whose accuracy and efficiency in the laboratory work were of invaluable help; to Dr. E. A. Rovenstine, Dr. Emmanuel M. Papper, and other members of the Department of Anesthesia of Bellevue Hospital; to Dr. Charles W. Lester and Dr. Gertrude B. Nicolson, who referred cases to us for study; to Miss Rae Morse, who prepared the manuscript; to Dr. Réjane Harvey and Dr. Irené Ferrer, who read it; to Dr. Domingo Gomez for his many helpful suggestions; to Dr. L. Emmett Holt and Dr. J. Burns Amberson, chiefs respectively of the Pediatric Service and the Chest Service of Bellevue Hospital; to Dr. Dickinson W. Richards, Jr., for his inspiring leadership and continuous encouragement; and finally to the Commonwealth Fund, which has supported the Cardio-Pulmonary Laboratory for many years and which undertook the publication of this monograph.

A. C.
J. S. B.
A. H.

October 1948

PART ONE

THE PHYSIOLOGICAL METHODS

THE physiological methods of the present study are based on the following three techniques (1) right heart catheterization (2) arterial puncture, and (3) collection of expired air for measuring the oxygen consumption All the measurements were made in the recumbent position and in the post absorptive state Occasionally a mild degree of physical activity was induced

The technique of right heart catheterization affords a means of sampling mixed venous blood in the right heart and the pulmonary artery In a given subject maintained in a steady state variations in oxygen content of blood samples taken in rapid succession from the right auricle the right ventricle the pulmonary artery and its two main branches are small Abnormal communications between these structures and the pulmonary vein the left auricle the left ventricle or the aorta result in contamination of mixed venous blood with highly oxygenated blood as the pressure conditions prevailing in the circulation favor blood shunts directed from the left cavities and their dependent vessels to the right cavities and the pulmonary artery Therefore the approximate location of the shunts can be determined by gas analysis and the volume of blood shunting can be calculated Moreover the presence of an abnormal communication may be revealed by the unusual routes eventually taken by the tip of the catheter as it is introduced into the heart Contamination of pulmonary venous blood by mixed venous blood can also be recognized by arterial blood gas analysis Such contamination occurs when the dynamic conditions prevailing in the heart in the presence of a cardiac malformation favor a shunt directed from the right cardiac cavities into the left cardiac cavities or the aorta

With simultaneous determination of the oxygen intake in the lungs and of the oxygen content in the heart and in a systemic artery the calculation of blood flow in the systemic and pulmonary circuits and of blood shunts through abnormal communication becomes a matter of applying simple formulae

In addition the two techniques of right heart catheterization and arterial puncture provide a means of recording blood pressures The patterns of the curves and the range of normal pressure variations in the right or left auricle the right or left ventricle the pulmonary artery, and the aorta and peripheral arteries are well defined Hence blood pressure recordings may serve two purposes (1) identification of the cardiac cavities or large vessels in which the tip of the catheter is placed (2) study of the effects of cardiac malformations upon the dynamics of the circulation

A well trained team and a well organized laboratory are essential for the successful application of the physiological methods The reliability of the results and the safety of the method depend upon meeting both these requirements

The team should consist of three physicians working as an integrated unit but each responsible for his part in the procedure and each constantly on the alert for changes in the patient s physiological status and for technical errors One member of the team thoroughly familiar with every aspect of the techniques and of the procedure should be in charge of the study so that the various steps may follow with a minimum of delay He directs the procedure and interrupts it immediately if he deems it advisable The laboratory should be organized with a group of technicians well trained in the rapid and accurate analysis of respiratory gases in air and in blood samples

The material presented in the following chapters may be of some help to investigators who are planning similar physiological studies It is the opinion of the authors however that a prolonged period of training with a team in a laboratory already in operation is the best means of acquiring a thorough knowledge of the method including its advantages as well as its pitfalls

Chapter I describes the equipment the techniques and the procedure employed in the course of the physiological studies and discusses the precautions to be observed in order to avoid undue risks and serious complications

Chapter II illustrates the various appearances of the catheter inside the heart and the large vessels, as shown by fluoroscopy.

Chapter III demonstrates the characteristic patterns of the blood pressure tracings, as they are recorded

Chapter IV presents the formulae used in connection with these studies for the calculation of the blood flow in the systemic or pulmonary artery and the intra and extracardiac blood shunts

Chapter I | EQUIPMENT, TECHNIQUES, AND PROCEDURE COMPLICATIONS

EQUIPMENT

The equipment used in the physiological studies includes venous catheters indwelling arterial needles apparatus for measuring oxygen consumption and for recording blood pressures an instrument set up for the introduction of the catheter and the needle and finally a large number of syringes for the collection of blood samples

Venous catheter The catheter* recommended is a modified ureteral type made of nylon with a smooth unwettable plastic covering flexible and radio opaque It is 100 cm long with only one opening at the tip and an airtight adaptor at the proximal end For convenience in manipulation a slight curve is provided in the distal 8 cm In the case of small children and infants a modified type with the curve starting 3 cm from the tip has been found useful A #6F or #7F catheter is preferable since pressure tracings are more satisfactory with these than with larger sized catheters and as a rule the withdrawal of blood samples is easy Sterilization is by boiling for 10 minutes in distilled water the catheter being then wrapped in a towel It should be sterilized some time before use because for a long period after heating it remains too flexible for easy manipulation Before introduction the catheter should always be carefully tested for surface imperfections and for leaks at the junction with the adaptor Immediately after use it should be rinsed with tap water for several minutes then filled with Haemo sol solution and allowed to soak in the same solution for 1 hour then rinsed again for 1 hour with tap water through a pressure connection *These precautions are indispensable in preventing the deposition of particles* of blood on the inner side of the woven fabric which might initiate a pyrogenic reaction in the course of the next catheterization

Indwelling arterial needle The needle† used for insertion in an artery is a modified Lindemann type in which a medium bevel #22 needle fits into a #19 outer sleeve with blunt tip The syringe adaptor is soldered about 1 5 cm down the #19 shaft the base of the shaft being allowed to project past the adaptor When a Luer lock syringe is attached for sampling this free segment goes directly into the end of the syringe and contamination from blood and blood clot lying in the fitting is thus avoided In the case of infants it is occasionally impossible to introduce an indwelling needle In this event a #20 needle with a short bevel is used for blood sampling and pressure recording but is not left in place

Instrument set up A sterile tray or truck is set up to carry the instruments and various sterile items required for skin incision isolation of the vein and closure of the incision They consist of two sheets and two large towels two towel clamps two small curved mosquito hemostats, two small straight mosquito hemostats a plain forceps a scalpel with a #11 blade a small scissors, a medicine glass for the novocain solution two #26 needles for local anesthesia two 5 cc. syringes a curved cutting edge needle with black silk for closure of the incision when necessary a small roll of black silk #0000 for ties several 3 way stopcocks and extra adaptors a large beaker to hold saline solution a 50 cc Luer lock adaptor syringe for testing the catheters for leaks a tourniquet Sterile gloves gowns sheets and infusion sets should be available

* Manufactured by the U S Catheter and Instrument Corporation *Glens Falls New York*
† Manufactured by Becton Dickinson *Rutherford New Jersey*

A second tray holds from fifteen to twenty 20 cc. syringes with Luer lock adaptors to be used in the sampling of blood These dry syringes are prepared by the introduction of a small globule of mercury, which fills the space between the rim of the plunger and the barrel and which is to be used in mixing the blood sample The sides of the syringe are then moistened with 6 drops of the following solution 600 mgm of sodium fluoride and 100 mgm of dried heparin in 15 cc of an 0 85 per cent sodium chloride solution The excess is expelled through the aperture of the syringe held in upright position the amount of solution left being too small to dilute the blood sample significantly If the fit of the plunger is not perfect, as sometimes happens its surface should be lightly greased with "Lubriseal" to avoid leaks and accumulation of air bubbles during sampling

Apparatus for the measurement of oxygen consumption The oxygen consumption may be determined by means of a standard metabolism apparatus, or by collecting samples of expired air in a Tissot spirometer or a Douglas bag and analyzing the gaseous content. In the case of infants and young children it is recommended that an anesthesia mask be used rather than a mouth piece or nose clip to connect the apparatus to the subject. A special mask* which has proved very satisfactory consists of a face piece of light weight plastic (moulded for form fitting) and a wide detachable cuff made of high grade inflatable rubber and built over a sponge rubber cushion

Apparatus for recording blood pressures For gross measurement of mean pressures an anaeroid Tycos manometer may be connected by a piece of rubber tubing to the catheter or the arterial needle A few cubic centimeters of saline solution introduced in the tubing in the part proximal to the catheter attachment, is used to minimize the chances of air embolism A saline manometer of the type used for the clinical measurement of venous pressure may be substituted for the anaeroid For accurate measurement and recording it is necessary to use fluid filled manometers of the Hamilton type or an electrical recording device In our work we have employed a special multiple blood pressure recording apparatus, designed and constructed by the Cambridge Instrument Company according to our specifications It consists of the following parts which are mounted on a mobile cabinet table (1) an electrocardiograph with camera string galvanometer time marker standardizing box, camera motor with 3 speed gear, (2) four fluid pressure Hamilton manometers with beryllium copper membranes of different sensitivities which can be connected through fluid filled lead tubing to the proximal end of the cardiac catheter and arterial needle, (3) a fluid system with two 3 way control valves and a pyrex bottle (2 000 cc capacity) the latter serving as a reservoir for a saline or sodium citrate solution and provided with a pressure bulb for applying pressure in the fluid system, (4) a calibrating mercury manometer and an air trap bottle, (5) an optical system for transmitting the deflection of the membranes to a recording camera with a light source and a reflecting mirror The optical system was designed in such a manner that there is no parallax between the simultaneous tracings of pressure and the electrocardiogram

TECHNIQUES AND PROCEDURE

Set up for study under fluoroscopic and electrocardiographic control The studies are carried out on a fluoroscopic table covered with a sponge rubber mattress. In order to avoid excessive radiation of the patient and of the members of the research team it is recommended that certain precautions be taken such as limitation of the field to be fluoroscoped the use of lead sheets as a protection against stray radiation, and the use of lead aprons and of a timing device

Throughout the entire period of study the electrocardiograph is connected with the subject and one member of the team follows the deflection of the string in Lead II in the reflecting mirror

* Manufactured by V Ray Bennett 8154 Beverly Boulevard Los Angeles California

for cardiac rate and irregularity, being particularly watchful during manipulation of the catheter within the heart

Anesthesia: In the case of children up to approximately 9 years of age catheterization is usually done under basal avertin anesthesia, given by a competent anesthetist, in order to maintain a steady state while obtaining multiple blood samples Premedication consists of scopolamine or atropine, occasionally with some barbital derivative Avertin is used in an initial dose of 90 to 110 mgm per kilogram of body weight and supplemented by doses of 20 to 40 mgm per kilogram if the anesthesia begins to wear off during the procedure In our studies the total dose was usually 140 mgm per kilogram For local anesthesia at the site of introduction of the catheter and of the arterial needle, a 2 per cent solution of novocain was used liberally

If it is thought that the child will cooperate and remain quiet throughout the procedure, local anesthesia at the sites mentioned above may be used—especially in the case of children of the older age group Occasionally an infant can be maintained in a quiet state without narcosis during the entire procedure The decision whether local anesthesia alone will suffice should be left to the physician best acquainted with the child's or infant's reactions

Technique of introducing the arterial indwelling needle: The special indwelling needle is placed in the brachial artery whenever possible Otherwise it is introduced in the femoral artery The site of the puncture and the deeper tissues are thoroughly infiltrated with a 2 per cent novocain solution The index finger is then placed over the infiltrated site and pressure is applied over the artery so that it stays in a given position while the underlying tissues are anesthetized It is advisable to infiltrate the tissues on either side of the artery as well as above and below the chosen route along which insertion of the arterial needle is to be made When the area has been anesthetized, the arterial needle containing the patent stylet is held in one hand while the index finger of the other applies sufficient pressure over the artery to fix its position but not to obliterate its pulsations The arterial needle is then inserted at a point just below the index finger and driven slowly through the skin and underlying tissues at an angle of about 45 degrees As progress is made in the direction of the pulsating vessel, the thumb is removed from the patent stylet at intervals in order to ascertain whether or not the artery has been punctured When puncture of the artery has been accomplished, blood will be seen to drip from the patent end of the stylet The flange of the needle is then grasped with one hand, the patent stylet with the other Coincident with the slow removal of the patent stylet, the arterial needle is "threaded" into the lumen of the vessel When the stylet has been removed, blood should spurt from the end of the needle The solid stylet is then placed in the needle, and stylet and needle together are threaded further into the vessel until a substantial portion of the needle lies within the lumen of the artery As soon as the needle is in place it should be taped securely to the skin and the arm or leg immobilized As a precaution against clotting in the needle, the solid stylet should be dipped in hydrogen peroxide before it is replaced During blood sampling, the stylet should stand in the peroxide

Technique of venous catheterization:

Choice of the site for introducing the catheter into the venous system Initially, a survey of the arms is made with a tourniquet applied If a vein going into the median basilic system can be felt or seen and is of sufficient size, it is selected as the site for introduction The left side is preferred because manipulation of the tip of the catheter inside the heart appears to be easier from this side but the right may also be used The choice of a vein draining into the cephalic system should be avoided since the acute angle at the subclavian vein can seldom be passed In the case of small children, or of those in whom no satisfactory arm vein can be found, the saphenous vein at the femoral area is used

Isolation of the vein·

Arm vein The arm is prepared by scrubbing with soap and water and then painting with tincture of merthiolate Sterile drapes are applied, so that the antecubital area is exposed in a sterile field The operator wears sterile gown and gloves

The area over the vein is infiltrated with a 2 per cent novocain solution A 5 mm incision is made parallel and just lateral to the course of the vein, which is exposed by blunt dissection with a small, curved hemostat and then elevated into the wound Two #0000 silk ligatures are passed under the vein and the distal one tied with a knot. The other is left untied and used as a sling to control back bleeding A transverse nick is made in the anterior wall of the vein with a #11 blade The proximal lip of the incision in the vein is grasped with a fine, straight hemostat and the lumen held open with it. The catheter is advanced into the vein after flow from the saline reservoir connected with it has been regulated at a rate of about 20 to 30 drops per minute

Saphenous vein The preparations are similar to those for the exposure of the arm vein *The femoral artery is palpated an inch below the inguinal ligament, and an area just* medial to it is infiltrated with 2 per cent novocain solution A 15 mm incision is made in the line of the vein, and the vein isolated by blunt dissection In infants this is a vessel from 2 to 3 mm in diameter *The vein is then handled in the same way as an arm vein,* the two ligatures being placed and the vein opened When the catheter is introduced, it can be felt to dip into the femoral vein, it is then advanced easily into the iliac veins

Manipulation of the catheter under fluoroscopic control After the catheter has been advanced 20 to 30 cm, further manipulation is done under fluoroscopic control This consists in advancing, rotating, and withdrawing the catheter until its tip is placed in the desired location When the catheter is introduced into an arm vein, some difficulty may occasionally be encountered in the axilla or the tip may be seen moving cephalad into the jugular vein If this occurs, the catheter is withdrawn, rotated, and advanced again with the neck flexed to the side, the arm abducted, and a slight traction exerted upon it If the catheter shows a repeated tendency to cross the midline into the opposite subclavian vein, advancing it at the time of inspiration will facilitate its entry into the superior vena cava The catheter should never be advanced against resistance Caution should be exercised to avoid buckling or the formation of a "U" When seen to be pointing in a desired direction, the catheter is advanced rapidly

To reach the right auricle, the tip is advanced until it lies about 2 or 3 cm above the level of the diaphragm To enter the right ventricle, the tip is rotated to point toward the left and then advanced On one such effort it will cross the midline Passage into the ventricle is checked by noting the pressure, with either a Hamilton or a Tycos manometer To enter the pulmonary conus, the catheter is carefully withdrawn and advanced, or rotated and advanced within the ventricle until the tip can be seen to point cephalad It is then advanced into the pulmonary conus and main branches of the pulmonary artery Occasionally in the case of infants, when the catheter has been introduced into the saphenous vein, a loop will form in the right auricle while an attempt is being made to advance the tip into the outflow tract of the right ventricle and the pulmonary conus The modified catheter with the curve near the tip was designed to remedy this difficulty As soon as the tip of the catheter has been placed inside the heart shadow, the saline flow from the reservoir may be reduced and adjusted to 5 to 10 drops per minute

Sequence of studies: The general plan of study consists in collecting blood samples and recording blood pressures in the heart and large vessels in rapid sequence In most instances the tip of the catheter is first advanced to one of the two main branches of the pulmonary artery, and

from there on withdrawn into the superior vena cava in successive stages during which blood samples are taken and pressures are recorded However unusual routes may be followed by the tip in various types of cardiac malformations In these cases fluoroscopic examination in various positions study of the pattern of the blood pressure tracings and gas analysis of the blood samples are essential in order to infer the location of the tip of the catheter In general blood samples and blood pressure tracings should be taken in quick succession in two communicating heart chambers or in a heart chamber and the communicating large vessel All blood samples should be withdrawn while the subject is maintained in a steady metabolic state If this steady state is interrupted some time should be allowed to elapse before resuming the collection of blood samples and the recording of blood pressures at the site where the last observations were made In a few cases of interventricular septal defect the oxygen content of the right auricle may be identical with that of the right ventricle in the quiet resting state and the shunt can be demonstrated only if the child becomes somewhat active or is induced to cry

If during the early part of the study the tip of the catheter has been introduced into a chamber of the left heart or into the pulmonary vein or the aorta blood samples and pressures should be taken immediately in these locations Otherwise the following order for sampling blood and recording pressures is recommended because it permits a rapid uninterrupted collection of data and a minimum of x ray exposure (1) one or if possible both branches of the pulmonary artery, (2) the truncus of the pulmonary artery (3) the outflow tract of the right ventricle (4) the mid portion or the apex of the right ventricle (5) the tricuspid area on the ventricular side, (6) the tricuspid area on the right auricular side (7) the mid right auricle (8) the superior vena cava (9) the inferior vena cava near its opening in the right auricle The blood sampling and pressure recording in the peripheral artery should be done simultaneously with similar sampling and recording in the right ventricle and if the opportunity presents itself in the aorta A protocol should be kept of the sequence of the study and a brief descriptive note of the exact location of the catheter should be dictated each time a physiological observation is made Occasionally x rays are taken in various positions when the location of the catheter cannot be easily identified or is unusual

Collection and handling of blood samples For the collection of blood samples the following procedure is recommended

A syringe containing a few cubic centimeters of a sterile saline solution and no air bubbles is adapted to the 3 way stopcock connecting the catheter to the saline reservoir Through proper manipulation of the stopcock and the syringe it is made certain that the circuit does not contain any air bubbles and that flow is free in the catheter Then suction is applied gently until blood flows back easily in the syringe *No forceful suction is used while sampling in the right ventricle since it will often cause ectopic beats* Then the Luer lock syringe used for sampling and prepared as described above is adapted to the 3 way stopcock and a blood sample of 5 to 10 cc obtained The sampling syringe is then disconnected and the 3 way stopcock and the catheter are immediately flushed with several cubic centimeters of a saline solution and the continuous drip started again Meanwhile the syringe with the blood sample is stoppered with a toothpick One small bubble of air is oftentimes present It can be expelled easily If the sample contains several bubbles especially in the form of foam it is discarded The syringe is rotated slowly while being taken to the laboratory Blood is transferred immediately under pressure into Ostwald Van Slyke pipette by way of a small (#23) gauge needle connected with the syringe and inserted into the tip of the pipette A small piece of rubber pierced by the needle makes possible an airtight seal The carbon dioxide and the oxygen content and the oxygen capacity are immediately measured in duplicate using the Van Slyke Neill apparatus The syringes containing the blood samples are

stored in an icebox for later drawing of samples if required In this case a period of 10 minutes should be allowed for rotation of the syringe before transfer of blood to the pipette

Recording of blood pressures The recording of blood pressure is done immediately before and after the collection of samples The free end of the lead tubings connecting with the manometer is fixed near the patient at a level corresponding to the plane of the heart 3 cm below the angle of Louis for intracardiac recordings, or at the level of the peripheral artery for intra arterial pressure recordings The free side arm of the 3 way stopcock connecting the catheter and the saline reservoir is flushed with fluid from the saline reservoir and then attached to the lead tubing The movements of the light beam are observed in the reflecting mirror in order to determine if artefacts are present or if the tracings are damped in which case the tip may be displaced or the catheter flushed with saline A short recording is taken the picture of the baseline being made at the beginning and at the end of each recording The film is immediately developed in an adjoining darkroom at the end of each step in the study

It is of great importance that the member of the research team in charge of the study should constantly be informed of the results of the gas analysis as they are obtained and be shown the tracings as they are developed In this manner the conduct of the study will be better controlled and plans altered according to findings As a rule, a continuous recording is made while the tip of the catheter is slowly moved from the region of the pulmonary artery into the outflow tract of the right ventricle under fluoroscopic control, and in cases in which a sudden systolic pressure increase is observed the point where this occurs is carefully noted This maneuver is essential in differentiating between stenosis at the pulmonary valve from stenosis of the infundibular area If a difference of pressure has been noted between the pulmonary artery and the ventricle attempts should be made to obtain repeated tracings while moving the catheter back and forth past the pulmonary valvular region

End of the procedure The catheter is withdrawn slowly but steadily If the arm vein was entered the tie applied to the vein at the beginning of the procedure is removed Pressure is used to control bleeding A small flamed adhesive strip is used to close the skin defect and a sterile dressing applied The arterial needle is withdrawn and immediate pressure on the vessel is maintained for at least 5 minutes and pulses below are carefully checked At any evidence of periarterial swelling pressure is reapplied If the saphenous vein was used it is tied near the confluence with the femoral vein penicillin is placed in the wound and the lips of the wound are sutured

COMPLICATIONS

Most complications observed during or after completion of cardiac catheterization are of a minor character Strict observance of details of technique given in the previous sections should help to avoid these It must always be kept in mind during the procedure that some risk is involved and that the safety of the patient dominates all other considerations The following is a brief discussion of the various complications related to the technique

Air embolism The accidental introduction of small air bubbles into the right cardiac cavities by way of the catheter is of no importance as long as there is no abnormal communication with the systemic circulation because these bubbles are probably effectively filtered out in the pulmonary capillaries In the presence of congenital malformations of the heart, associated with hemodynamic conditions favoring right to left blood shunts air emboli may be thrown into the systemic circulation if precautions against the introduction of air bubbles into the catheter are

not taken Embolization of one or several cerebral arteries is one of the most serious hazards of using the technique in this type of defect. However, in his studies of several hundred cases of intracardiac communications, Bing suspected it in two cases only In no instance did it result in more than transient neurological symptoms or signs

Venous and intracardiac thrombosis: The trauma incident to the incision of the vein and the introduction of the catheter often causes a limited thrombosis This usually extends less than 5 cm upward from the site of the incision In order to maintain a free blood flow in the main brachial vein it is therefore very important to choose as the site of introduction of the catheter a branch of the basilic venous system which is distal to and at a distance of at least 5 cm from the last confluence of veins of this system Potentially, catheterization of the saphenous vein offers a greater risk of thrombosis of larger veins—namely, the femoral, with the attendant danger of pulmonary emboli In order to minimize this risk, the saphenous vein should be tied off as close to the femoral vein as possible at the end of the procedure

Intracardiac thrombosis, associated with traumatic lesions of the endocardium, is another potential hazard to be reckoned with Goodale, Lubin, Eckenhoff, Hafkenschiel, Durlacher, Landing and Banfield have described lesions following the prolonged introduction of catheters into the hearts of dogs * With one exception, however, such lesions have not been revealed in the pathological material gathered by many groups of investigators using the technique of cardiac catheterization in man Only one autopsy report, by Johnson, Wollin, and Ross, is suggestive of an endocardial lesion In the case of a young child with polycythemia and a congenital cardiac defect, who died one month after catheterization, a thrombus was found in the right auricle, inferior vena cava and iliac and renal veins It is not clear from the description given whether the thrombus originated at the site of introduction of the catheter in the saphenous vein, or at the base of a leaflet of the tricuspid valve to which it was attached, or in both locations Whether the hazard of venous thrombosis in patients with polycythemia is serious enough to warrant prophylactic anticoagulant therapy after catheterization is an important question Because of our limited experience with this group, we are not in a position to give a documented answer

Rhythm disturbances: The development of rhythm disturbances during cardiac catheterization is by far the most important complication to consider

Premature contractions are often observed during the manipulation of the catheter inside the heart These ectopic beats usually occur when the tip of the catheter is being passed through the tricuspid valve, or lies against the ventricular septum in the region of the outflow tract, or when suction is applied in sampling blood in the right ventricle Ectopic beats are easily identified by watching the deflection of the galvanometer string. Changing the position of the catheter causes these beats to disappear

Runs of ventricular tachycardia have been encountered on a very few occasions when the cause of abnormal stimulation was not removed In our studies of congenital malformations of the heart we observed one patient with an interventricular septal defect in whom the introduction of the tip of the catheter through the defect stimulated many ectopic beats of bizarre electrical configuration and short runs of ventricular tachycardia These ceased upon withdrawal of the catheter into the right auricle, but the episode was serious enough to cause signs of mild vascular collapse One can easily visualize the possible development of ventricular fibrillation in the course of such an attack of ventricular tachycardia This fatal complication has never been observed, as far as we know, by any of the groups investigating congenital heart disease Death

* Coronary sinus catheterization technique for studying coronary blood flow and myocardial metabolism in vivo Proc Soc Exper Biol of Med *66* 571, 1947

is known to have occurred suddenly, however, in the case of two adult cardiacs, one of them with a long history of angina pectoris, but in neither instance was the catheterization performed with electrocardiographic control It may be hypothesized that early recognition of ectopic beats or of runs of ventricular tachycardia would have led to withdrawal of the catheter, thus possibly preventing these accidents

A paroxysm of auricular tachycardia has been reported by Johnson et al in a case of interauricular septal defect. We have observed two such episodes in the course of our studies In one case, in which previous attacks of a similar nature had been noted, the paroxysm ceased spontaneously after a short time, in the other it stopped immediately after displacement of the catheter

Blood loss: The volume of blood withdrawn during the procedure is not less than 100 cc. This amount constitutes a fairly large proportion of the total circulating blood volume in young children and infants, the loss of which is likely to upset the steady state of the circulation In such cases it is therefore important to estimate the volume of blood withdrawn during each step of the procedure and to replace it with an equivalent amount of donor blood

Chapter II ‖ X-RAY OBSERVATIONS DURING CATHETERIZATION

The following figures illustrate the various locations of the tip of the catheter as seen on the fluoroscopic screen or recorded on x-ray films during the studies.

The first seven figures show the catheter with the tip located successively in (1) the right auricle. (2) the right ventricle pointing toward the apex, (3) the outflow tract of the right ventricle, (4) the truncus of the pulmonary artery near the valve and (5) near the bifurcation, (6) the left pulmonary artery, (7) the right pulmonary artery.



FIG. 1

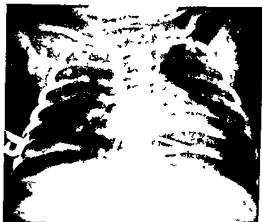


FIG. 2

Figure 1: Catheter inserted in the right saphenous vein. The tip is located in the right auricle.

Figure 2: Catheter inserted in the right saphenous vein. The tip has been advanced through the tricuspid valve into the right ventricle and is pointing toward the apex.



FIG. 3

Figure 3: Catheter inserted in the left median basilic vein (Case 1). The tip has been advanced through the tricuspid valve into the right ventricle, pointing cephalad, and is located in the outflow tract. As premature ventricular contractions are apt to occur in this location, the catheter should never be left here without electrocardiographic control; and even with such control it should preferably be left for only a short time.


FIG 4


FIG 5

Figure 4: Catheter inserted in the left median basilic vein (Case 2) The tip is just on the arterial side of the pulmonary valve

Figure 5: Same case as Figure 4 The tip is near the bifurcation of the main pulmonary artery


FIG 6


FIG 7

Figure 6: Catheter inserted in the left median basilic vein (Case 13) The tip is in the left pulmonary artery

Figure 7: Catheter inserted in the right saphenous vein (Case 9). The tip is in the right pulmonary artery In this position the tip of the catheter can often be seen moving back and forth several centimeters

Placement of the tip of the catheter in the various locations illustrated in the above seven figures is attempted routinely It is usually easy unless tricuspid insufficiency or pulmonic atresia is present

The remaining figures picture some of the unusual paths taken by the catheter in septal defects and complex cardiac anomalies.

In the next group of four figures the catheter tip has been introduced through an interauricular septal defect into the left auricle and then into one of the pulmonary veins or has been moved from the right auricle directly into a pulmonary vein



FIG 8



FIG 9

Figure 8: Catheter inserted in the right saphenous vein (Case 9). The tip has been introduced through an interauricular septal defect into the left auricle. In comparing with Figure 2, where the catheter is passing through the tricuspid valve into the right ventricle, note that the curve here is higher and less acute

Figure 9: Catheter inserted in the right saphenous vein (Case 8). The tip, introduced into the left auricle through an interauricular septal defect, has been placed in a left pulmonary vein.



FIG. 10



FIG 11

Figure 10: Catheter inserted in the right saphenous vein The tip, introduced into the left auricle through an interauricular septal defect, has been placed in a left superior pulmonary vein The exact location was ascertained by study of the pattern of the blood pressure tracing and by blood gas analysis Otherwise, it could be mistaken for the left innominate vein or even a left pulmonary artery (compare with Fig. 6).

Figure 11: Catheter inserted in the right saphenous vein (Case 9). The tip has been introduced from the right auricle directly into a right pulmonary vein Without the confirmation of the blood gas analysis, it could be assumed that the tip was in the right hemi-azygos vein.

In the following group of five pictures the catheter tip has been introduced into the left ventricle through an interventricular septal defect or into an overriding aorta.



FIG. 12



FIG. 13

Figure 12: Catheter inserted in the right saphenous vein (Case 12). The tip has been introduced through the tricuspid valve into the right ventricle and through an interventricular septal defect into the left ventricle. The location was confirmed by fluoroscopy in the oblique position (see Fig. 13), by recording the blood pressure (see Fig. 25), and by analysis of blood samples With the catheter crossing the ventricular septum, the dangers of multiple premature contractions and ventricular tachycardia must constantly be kept in mind. To introduce the catheter into the left ventricle is not advisable.

Figure 13: The same case in the left anterior-oblique position.



FIG. 14



FIG. 15

Figure 14: Catheter inserted in the right median basilic vein. The tip has been introduced through the right ventricle into an overriding aorta with a left aortic arch. The position was confirmed by blood pressure tracings (see Fig. 27) and blood gas analysis.

Figure 15: The same case in the left anterior-oblique position.



FIG 16

Figure 16: Catheter inserted in the right median basilic vein (Case 16). The tip has been introduced through the right ventricle into an overriding aorta with a right aortic arch

The next four figures illustrate some of the unusual x-ray aspects observed in a case of dextro cardia (Case 17) with multiple congenital defects of the heart.



FIG. 17



FIG 18

Figure 17: Catheter inserted in a right median basilic vein The tip was then introduced successively in a right superior vena cava, an auricular chamber with low blood pressure, and a left superior vena cava.

Figure 18: Catheter reinserted in the same right median basilic vein and seen to be crossing into a left superior vena cava The tip was later introduced into a cardiac chamber with low blood pressure, then into a chamber with high blood pressure, the pattern of which was characteristic of the ventricle; and was finally placed in the pulmonary artery, identified only by the blood pressure tracings.


FIG 19

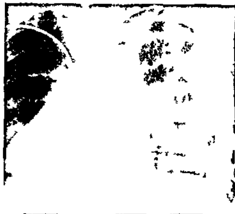

FIG 20

Figure 19: The tip of the catheter is in a cardiac chamber with low blood pressure (auricular pattern)

Figure 20· Here the tip is in a cardiac chamber with high blood pressure (ventricular pattern)

Chapter III — CHARACTERISTIC PATTERNS OF THE BLOOD PRESSURE TRACINGS

Determination of the location of the tip of the catheter is considerably helped by recording the blood pressure tracings and by immediately developing and examining the films. By watching the deflection of the manometer's light beam on the reflecting mirror of the recording apparatus, it is generally possible to ascertain, once the catheter is connected with the manometer, whether the tip of the catheter is in the right auricle, the right ventricle, or the pulmonary artery, as the patterns of the curves are quite distinct The cyclic variations in the left auricle are of greater amplitude than those in the right auricle, and it requires a close inspection of the record to decide whether the tip of the catheter is in the left auricle or within the tricuspid valve area of the right ventricle. It is the timing of the large upward deflection, in relation to the P and QRS waves of the electrocardiogram. that provides the best means for differentiation. The blood pressure tracings respectively in the two ventricles and in the pulmonary artery and aorta are similar in form. They can only be differentiated by the height of pressures if normal conditions prevail in the right heart, as the systolic pressure in the left heart is about five times higher than in the right. Therefore, if the blood pressures are extremely elevated in the right heart, differentiation is based upon fluoroscopic observations and the results of the blood gas analysis.

The most characteristic patterns of blood pressures as recorded in the right auricle, the left auricle, the right ventricle, the left ventricle, the pulmonary artery, and the aorta are illustrated in Figures 21 to 26.

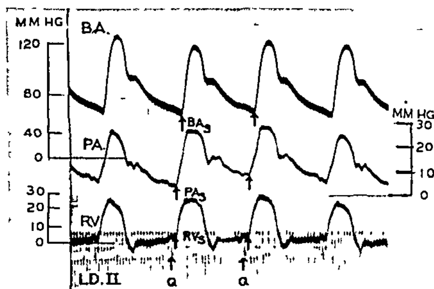


FIG. 21

Figure 21: Normal blood pressure patterns in the brachial artery, the pulmonary artery, and the right ventricle. In the pulmonary artery and right ventricular tracings the upward deflection starts after the beginning of the QRS complex. The slope of the downward deflection is very steep in the right ventricle, corresponding to the isometric relaxation preceding the plateau-like diastolic curve, and contrasts with the progressively downward diastolic curve in the pulmonary artery. As seen from the scale, the systolic pressure is less than 30 mm. Hg in the right ventricle and in the pulmonary artery.

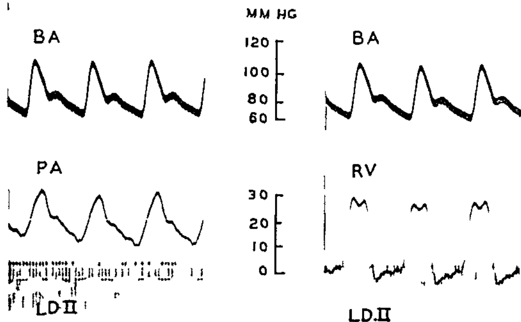


Figure 22· Blood pressure tracings in the brachial artery, the pulmonary artery, and the right ventricle in a case of interauricular septal defect. The blood pressure recordings were taken through a double lumen catheter without change in its position and in rapid succession In spite of a large increase in pulmonary blood flow, the blood pressures in the pulmonary circulation were normal This corresponds to similar observations made in normal subjects, indicating that a threefold increase in pulmonary blood flow does not cause any increase in pulmonary arterial pressure The portion of the ascending slope corresponding to ejection in the right ventricle is much steeper than the analogous portion in the pulmonary artery This difference in slope may be due in part to physical characteristics of the pulmonary arterial system (deformability of the large vessels, resistance in the small vessels, blood mass, etc), in part to the velocity of ejection and probably in some part to the recording of pressure through the catheter

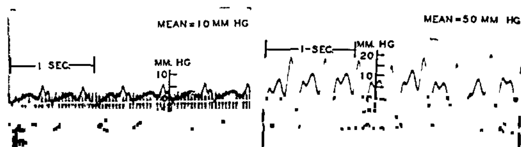


Figure 23: Blood pressure pattern in both auricles Tracings taken in the right and left auricles of an infant of 5 months with an interauricular septal defect. To the left, the tracing taken in the right auricle the main upward deflection (A) starts after the P wave of the electrocardiogram and before the QRS It corresponds to the auricular systole The downward slope of the A wave is interrupted by a plateau, followed by a dip both corresponding to isometric contraction and beginning ejection in the right ventricle. The pressure variations during the entire cycle are small The normal mean pressure varies within narrow range, with an upper limit of about 5

mm. Hg. To the right, the tracing taken in the left auricle: the main upward deflection, starting after the P wave but before the QRS complex, corresponds to the auricular systole. The downward stroke of the A wave merges with the marked dip corresponding to the left ventricular isometric contraction and ejection The pressure rises immediately following this dip during the remainder of the ventricular systole. It drops markedly again at the time of the opening of the mitral valve The height of the deflection corresponding to the auricular systole is much greater in the left than in the right auricle. The mean pressure in the left auricle is from 2 to 4 mm. Hg higher than in the right auricle.



FIG. 24

Figure 24: Blood pressure pattern simulating left auricular tracings. Tracing taken with the tip of the catheter in the tricuspid valve area. The upward deflection corresponds to the ventricular systole. It starts after the beginning of the QRS complex. The whole curve is an artefact. (For comparison see Fig. 23, right curve)

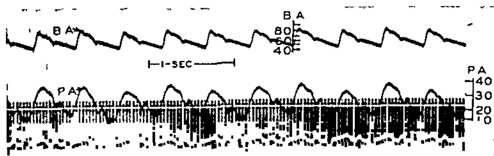


FIG. 25A

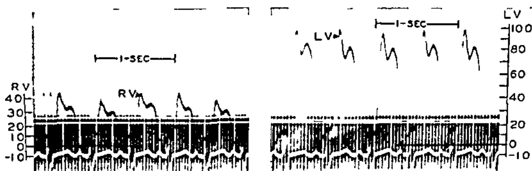


FIG. 25B

Figure 25: Blood pressure patterns in the left and right ventricles and in the pulmonary artery. Tracings taken in the case of a child of 3 years with an interventricular septal defect (Case 12). The left ventricular blood pressure tracing is identified by the large systolic deflection and the higher diastolic level Otherwise, in form and in timing it is identical with that of the right ventricle.

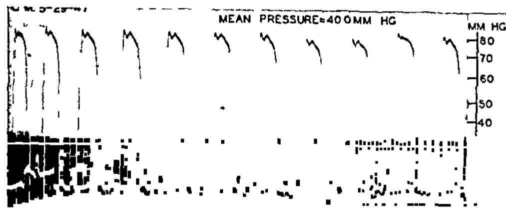


FIG 26

Figure 26: Blood pressure pattern in the right ventricle Tracing taken in the case of a child of 6 years with an interauricular septal defect and right ventricular hypertension It resembles in all respects a left ventricular tracing (see Fig 25) Right ventricular hypertension not associated with pulmonary stenosis, or overriding of the aorta, or transposition of the large vessels reflects either an increase in pulmonary vascular resistance, or hypertension in the left auricle and pul monary veins, or a combination of both

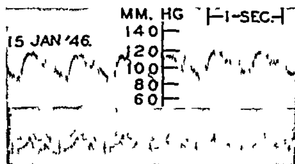
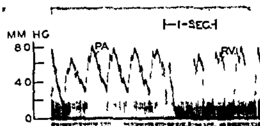


FIG 27

Figure 27: Blood pressure pattern in the aorta Tracing taken in the case of a child of 12 years with *a tetralogy of Fallot* The pattern is that of a central artery Such tracings do not differ in form from those taken in the pulmonary artery Therefore in instances of transposition of the large vessels analysis of the tracings cannot be of diagnostic value

Aside from helping to determine the location of the tip of the catheter and to measure blood pressures in various cardiac cavities and large vessels, blood pressure recordings are of great value in the diagnosis of pulmonary stenosis The next three figures show typical patterns of blood pressure tracings taken as the catheter is being withdrawn from the pulmonary artery into the right ventricle



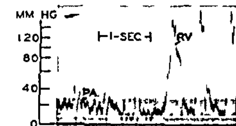
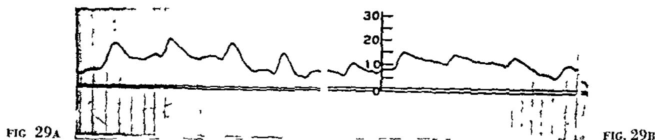


FIG. 28

A WITHOUT PULMONARY STENOSIS B WITH PULMONARY STENOSIS

Figure 28: Blood pressure tracings taken during withdrawal of the catheter from the pulmonary artery into the right ventricle (Case 10) To the left (A), typical finding in a subject with out pulmonary stenosis but with pulmonary hypertension The systolic pressures are identical in the pulmonary artery and the right ventricle The slope of the downward curve and the dias-

tolic level are different. To the right (B), typical finding in a case of tetralogy of Fallot with pulmonary stenosis. The mean pressure in the pulmonary artery is very low. The pressure rises suddenly as the tip reaches the right ventricular outflow tract. Many vibrations are set up by the motion of the catheter during withdrawal.



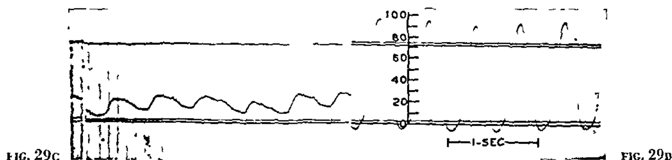


Figure 29: Blood pressure tracing taken during withdrawal of the catheter from the pulmonary artery into the right ventricle in a case of stenosis of the infundibulum.

The tracings were taken with the tip successively in the left branch of the pulmonary artery (A), the pulmonary truncus (B), the infundibular area of the right ventricle (C), and finally the right ventricle (D). The arterial pattern recognizable in the first two is less distinct in the infundibulum. When the tip reached a point in the ventricle very near the tricuspid valve, the pressure rose markedly and the blood pressure pattern became that of a right ventricle with hypertension. The exact location of the tip, as it is seen on the fluoroscopic screen when such a pressure change occurs, is an important landmark in differentiating between valvular and infundibular stenosis.

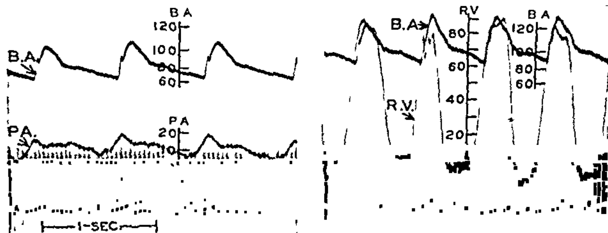


Figure 30: Blood pressure tracings taken with the catheter successively in the pulmonary artery and the right ventricle (Case 1). The same observations were made as in the previous case. The tracing in the infundibular region has not been reproduced.

In the case of some patients with marked dilatation of the pulmonary artery of congenital origin, but without other malformations, a difference in systolic pressure exists between the pulmonary artery and the right ventricle, although the blood pressures in the latter may be within normal limits. A tracing taken in such a case is illustrated in the next figure.

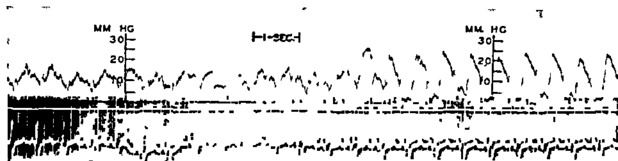


FIG 31A FIG 31B

Figure 31: Blood pressure tracings taken in a case of idiopathic dilatation of the pulmonary artery (Case 2).

The mean pressure in the pulmonary artery (A) is normal, the pulse pressure small. The systolic pressure in the right ventricle (B), although nearly 10 mm Hg higher than in the pulmonary artery, is normal. The same observation was made several times by moving the tip of the catheter back and forth in the valvular region. The abnormal expansibility of the dilated pulmonary artery, with its thin wall, is probably the chief cause of these pressure differences.

The blood pressure recordings from a peripheral artery may reveal abnormalities in the region of the aortic valve or further along the course of the aorta. Such a tracing is illustrated in the next figure.



FIG. 32

Figure 32: Blood pressure tracing taken in a case of subaortic stenosis (Case 11). The plateau-like systole and the small pulse pressure are consistent with such a condition.

Chapter IV | FORMULAE FOR THE CALCULATION OF SYSTEMIC AND PULMONARY BLOOD FLOW AND OF BLOOD SHUNTS

SYMBOLS

The data used for the calculation of the systemic or pulmonary blood flow and of various blood shunts are represented in the formulae by the following symbols

Oxygen content in blood samples: The letter "C" (for concentration) with a subscript indicating the location is used as a symbol to represent the oxygen content in the sample expressed in cubic centimeters per liter of blood

C_{PA} = Oxygen concentration in the pulmonary artery
C_{RV} = Oxygen concentration in the right ventricle
C_{RA} = Oxygen concentration in the right auricle

In individuals without an abnormal communication between the left and the right heart, the concentration of oxygen in these three chambers or vessels usually checks within 3 cc per liter of blood It differs exceptionally by as much as 6 cc per liter of blood

C_{SVC} = Oxygen concentration in the superior vena cava
C_{IVC} = Oxygen concentration in the inferior vena cava
C_{VC} = Oxygen concentration in the venae cavae, which by definition is derived from the following formula $\frac{C_{SVC} + C_{IVC}}{2}$

C_{VC} is a calculated figure, as a rule not significantly different from C_{RA} in individuals without congenital heart defects Occasionally the two columns of blood joining in the right auricle may remain separate as far as the right ventricle (streamline flow)

C_{Ao} = Oxygen concentration in the aorta or in any peripheral artery
C_{LV} = Oxygen concentration in the left ventricle
C_{LA} = Oxygen concentration in the left auricle
C_{PV} = Oxygen concentration in a pulmonary vein

Unless there is a shunt from right to left, the concentration of oxygen in the peripheral artery is identical with that in the aorta the left ventricle, the left auricle, and the pulmonary veins The normal oxyhemoglobin saturation in the arterial blood is 96 per cent or better, except in cases of interauricular septal defect, where C_{LA} may be measured by direct sampling the assumption is made in the calculation of pulmonary blood flow that C_{LA} corresponds to a blood 96 per cent saturated with oxygen

Oxygen consumption = (O_2) The oxygen consumption is expressed in cubic centimeters per minute It is measured under basal metabolic conditions and in a steady state The oxygen consumption in the tissues is equal to the oxygen intake in the lungs under these conditions

Systemic and pulmonary blood flow: The systemic and the pulmonary blood flow are expressed in liters per minute and are represented by the following symbols

Q_S = Systemic blood flow
Q_{PA} = Blood flow in the pulmonary artery
Q_{PC} = Pulmonary capillary blood flow

The pulmonary blood flow is equal to the pulmonary capillary blood flow in all instances, except when anastomoses to the pulmonary arterial system are supplied by the aortic system beyond the pulmonary artery or its main branches In these instances Q_{PC} is greater than Q_{PA}

Blood shunts: Blood shunts between the right heart and the pulmonary artery, and the left heart *and the aorta, expressed in liters per minute are represented* by the following symbols, which indicate the direction and the location of the shunt

X = Right to left shunt of blood
Y = *Left to right shunt of blood*
Y'_1 = Shunt of blood from pulmonary veins or the left auricle into the right auricle
Y_2 = Shunt of blood from the left into the right ventricle
Y_3 = Shunt of blood from the aorta into the pulmonary artery
R = Blood flow in regurgitation through a cardiac valvular region, usually the pulmonic valve

FORMULAE

The following formulae have been derived for the various calculations of systemic blood flow, pulmonary blood flow, and blood shunts

Estimated systemic blood flow

$$Q_s = \frac{O_2}{C_{Ao} - C_{RA} \text{ or } C_{VC}} \qquad (1)$$

Estimated pulmonary blood flow: There are two cases to consider

First case $Q_{PA} = Q_{PC}$, *then*

$$Q_{PA} = \frac{O_2}{C_{LA} - C_{PA}} \qquad (2)$$

This formula can only be used if no anastomoses are supplied by the aortic system to the pulmonary circulation beyond the point of sampling It is essentially the same formula as formula 1 because oxygen consumption in the tissues is equal to oxygen intake by the lungs, C_{LA} is equal to C_{Ao} and C_{PA} is equal to C_{RA}

Second case Q_{PA} is less than Q_{PC}

When it is suspected that anastomoses are supplied by the aortic system to the pulmonary circulation beyond the point of collection of blood samples in the pulmonary artery (C_{PA}), then Q_{PC} cannot be measured accurately, but it may be estimated as being between the two following limits

$$\frac{O_2}{C_{LA} - C_{PA}} \leq Q_{PC} \leqq \frac{O_2}{C_{LA} - C_{Ao}} \qquad (3)$$

In the left part of the formula it is assumed *that the pulmonary capillary flow is supplied entirely* by the pulmonary artery, in the right part, it is assumed that the capillary flow is supplied entirely by the aortic system Obviously the pulmonary capillary blood flow lies somewhere between the limits thus defined An estimate of this value may be clinically useful in cases where it is important to decide whether the pulmonary arterial blood flow is reduced

Blood shunts There are several cases to consider, depending upon the oxygen concentration of the blood in various samples

First case C_{RA} is greater than C_{VC}

Mixture in the right auricle of blood shunting from the left auricle or pulmonary vein with blood flowing from the venae cavae is suspected here Another interpretation would be an interventricular septal defect with tricuspid insufficiency Clinical examination and the special pattern of the blood pressure tracing in the right auricle would help in recognizing such a combination The formula used to calculate the shunt is as follows

$$Y_1 = Q_S\left(\frac{C_{RA} - C_{VC}}{C_{LA} - C_{RA}}\right) \qquad (4)$$

Second case C_{RV} is greater than C_{RA}

Mixture in the right ventricle of left ventricular blood shunting through an interventricular septal defect with blood flowing from the right auricle is suspected The difference may be present in all three of the samples from the right ventricle or only in the sample from the outflow tract. The formula used to calculate this shunt is

$$Y = Q_S\left(\frac{C_{RV} - C_{RA}}{C_{LV} - C_{RV}}\right) \qquad (5)$$

If in addition C_{RA} is greater than C_{VC} the following formula is used

$$Y = (Q_S + Y_1)\left(\frac{C_{RV} - C_{RA}}{C_{LV} - C_{RV}}\right) \qquad (6)$$

Third case C_{PA} is greater than C_{RV}

In this case mixture in the pulmonary artery of blood shunting from the aorta through a patent ductus arteriosus or an aortico pulmonary septal defect with blood flowing from the right ventricle is suspected Large differences in the various blood specimens sampled in the truncus and the main branches of the pulmonary artery depend upon (a) the location of the tip of the catheter with regard to the column of blood coming from the aorta and (b) the degree of mixing which takes place The choice of a figure to use in the calculation may cause variation in the estimated blood shunts over a considerable range The formula used to calculate this shunt is

$$Y_3 = Q_S\left(\frac{C_{PA} - C_{RV}}{C_{Ao} - C_{PA}}\right) \qquad (7)$$

Fourth case C_{PA} greater than C_{RV} and C_{RV} greater than C_{RA}

There is a choice between two formulae when it is found that in addition to an aortico pulmonary shunt C_{RV} is greater than C_{RA} The choice depends on whether the clinical findings support the diagnosis of (a) an aortico pulmonary shunt associated with an interventricular septal defect or (b) an aortico pulmonary shunt associated with pulmonic incompetence

In the first instance the interventricular shunt is calculated from formula 5 and the aortico pulmonary shunt from the following formula

$$Y_3 = (Q_S + Y)\left(\frac{C_{PA} - C_{RV}}{C_{Ao} - C_{PA}}\right) \qquad (8)$$

In the second instance the aortico pulmonary shunt is calculated from formula 7 and the volume of pulmonic regurgitation is calculated from the following formula

$$R = Q_S \left(\frac{C_{RV} - C_{RA}}{C_{PA} - C_{RV}}\right) \quad (9)$$

If an interventricular septal defect is associated with a patent ductus arteriosus and pulmonic incompetence, it is impossible to measure separately the flow of blood shunting from the left to *the right ventricle* and regurgitating through the incompetent valve

It should be noted that, in the absence of an aortico pulmonary shunt, the diagnosis of pulmonic regurgitation cannot be established on the basis of blood gas analyses

Fifth case The oxyhemoglobin saturation of the arterial blood is reduced below the normal value of 96 per cent

A right to left shunt is suspected, and the blood flow through the shunt is calculated from the following formula

$$X = Q_S \left(\frac{C_{LA} - C_{Ao}}{C_{LA} - C_{VC}}\right) \quad (10)$$

This shunt most commonly occurs between the two auricles or between the right ventricle and an overriding aorta Very rarely it may take place between the pulmonary artery and the aorta as a result of partial reversal of blood flow through a patent ductus arteriosus In this instance formula 10 is not applicable

In the case of an interventricular septal defect combined with overriding of the aorta in which there is a double blood shunt from left to right and from right to left, formulae 5 and 10 measure respectively (a) the volume of blood of left auricular composition which flows into the pulmonary artery and (b) the volume of blood of right auricular composition which flows into the aorta The approximate volume of the columns of blood circulating in each direction, across the defect and becoming intermixed in a manner impossible to predict may be calculated if equation 5 is expressed as follows

$$Y' = Q_S \frac{(C_{PA} - C_{RA}) \quad (C_{AO} - C_{RA})}{(C_{LA} - C_{PA}) \quad (C_{LA} \quad C_{RA})} \quad (5a)$$

and if $X = Q_S + Y'_2 - Q_{PA}$ (11)

In some instances a special technique is required to differentiate the oxygen unsaturation of the arterial blood, produced by a shunt of mixed venous blood from that due to inadequate oxygenation of capillary blood in the lungs This technique consists in collecting arterial blood samples while the subject is breathing pure oxygen If the shunt is 25 per cent or more, the arterial blood fails to become fully saturated with oxygen

PART TWO

ILLUSTRATIVE CASES

A uniform plan has been followed in the presentation of the seventeen individual cases selected for discussion The data and comments are given in the following order

1 The clinical data, with emphasis on the cardiac signs and the x ray findings

2 The clinical diagnosis, prior to the physiological studies

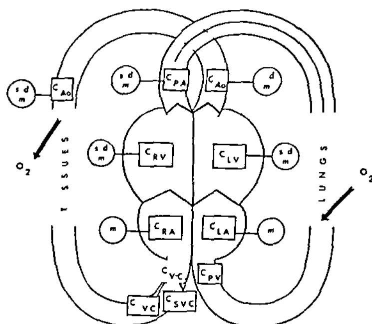
3 The physiological data, presented in a schematic form (see the symbols listed on pages 26–27, the diagram illustrated below, and the key on the opposite page)

4 The calculations of blood flow and blood shunts computed on the basis of the oxygen content in the venous blood samples and of the oxygen consumption

5 Comments concerning the validity and significance of the observations made in the course of the physiological studies

6 The follow up history, including a brief description of operative or autopsy findings when available

7 A general comment combining and correlating all the observations



SCHEME OF THE DIAGRAMS

KEY TO THE DIAGRAMS

Blood Oxygen Content

C cc/lit of blood = observed

cc/lit. of blood = assumed or derived

Blood Pressures in mm Hg

s/d m — s = systolic, d = diastolic, m = mean

Valvular Lesions

Dilatation
Competent valve
Stenosis

Incompetent valve
Stenosis

Blood Oxygen Saturation

0 to 20%

21 to 40%

41 to 60%

61 to 70%

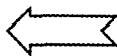
71 to 80%

81 to 90%

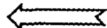
90% and over

Shunts—Direction and Volume

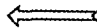
Shunt equal to or greater than systemic flow



Shunt between 33% and 100% of systemic flow

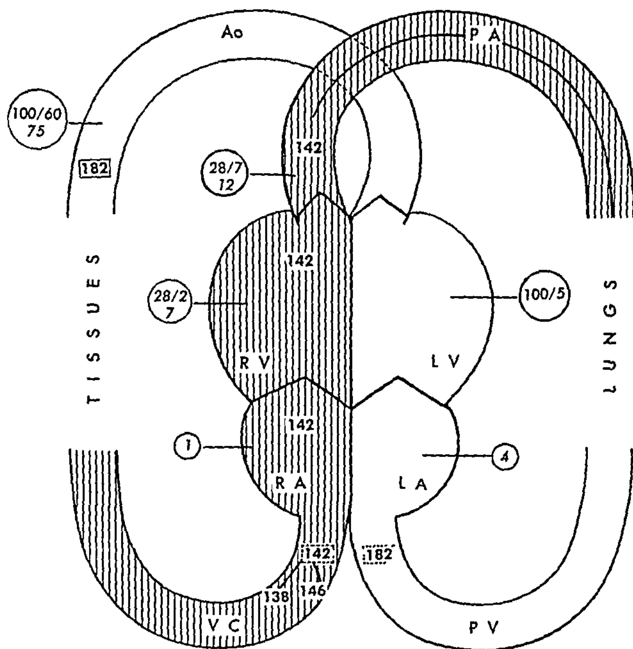


Shunt less than 33% of systemic flow



APPROXIMATE HEMODYNAMIC DATA

IN A NORMAL CHILD—AGE 3 YEARS, WEIGHT 14 KG.



Arterial blood oxygen capacity . . = 192 cc/lit.
Arterial blood oxygen saturation = 96%
Oxygen consumption . . . at rest = 100 cc/min.

ESTIMATED BLOOD FLOW

Systemic (Q_S) . . . . . . 2.50 lit/min.
Pulmonary (Q_{PA}) . . . 2.50 lit/min.

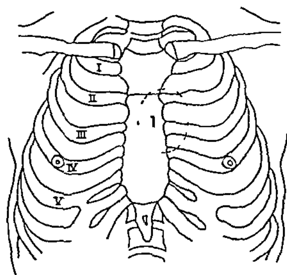
Case 1 ‖ INFUNDIBULAR PULMONARY STENOSIS POST-STENOTIC DILATATION OF PULMONARY ARTERY

Clinical data: Age 8 years Weight 25 4 Kg (56 lbs) F

History: This child was somewhat slender but otherwise well developed No clubbing dyspnea, or easy fatigue had been noted

Physical examination: There was no cyanosis or dyspnea on considerable exercise

HEART



Size Slightly enlarged to left

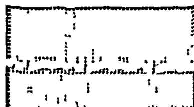
Thrill Coarse systolic close to sternum at II left interspace

Sounds P_2 faint

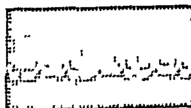
Murmurs 1 Long, loud, rather coarse systolic over upper left precordium, maximum at III left interspace at sternal edge, did not transmit well toward left clavicle

Arterial blood pressure 112/72 mm Hg, after exercise, 118/76 mm Hg

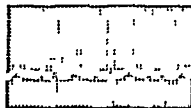
ELECTROCARDIOGRAM



LEAD I

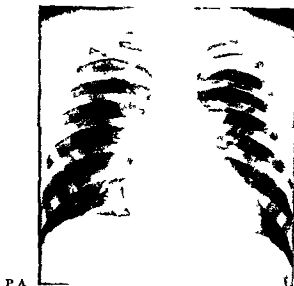


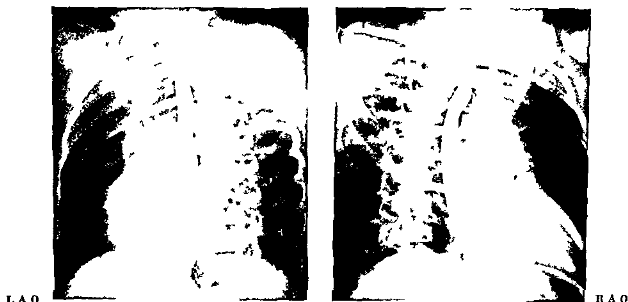
LEAD II



LEAD III

Slight right axis deviation Normal conduction time Slight notching of P waves

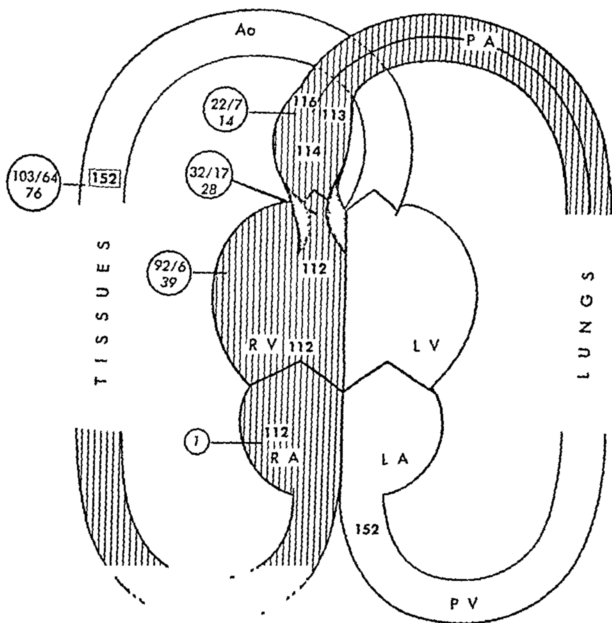




There was slight transverse enlargement of the heart with the apex lifted well above the diaphragm In the region of the pulmonary arc there was a prominent rounded shadow which showed very little pulsation A slight concavity was present immediately below this shadow In the right anterior oblique view there was some flattening in the region of the outflow area of the right ventricle with prominence of the pulmonary artery above In the left anterior oblique view the right ventricle showed moderate enlargement but the left ventricle did not appear enlarged There was no increase in the vascular markings in the lung fields

CLINICAL DIAGNOSIS

Pulmonary stenosis with post stenotic dilatation of the pulmonary artery The presence of other anomalies which are frequently associated with pulmonary stenosis, such as an interventricular septal defect and overriding of the aorta in various degrees, was considered unlikely on the basis of the physical signs and the size and shape of the heart as shown by x ray, and in view of the lack of any suggestion of cyanosis after considerable exercise



Arterial blood oxygen capacity = 162 cc/lit
Arterial blood oxygen saturation at rest = 95%
Arterial blood oxygen saturation following mild exercise = 94%
Oxygen consumption at rest = 186 cc/min

ESTIMATED BLOOD FLOW

Systemic (Q_s) 4 78 lit/min
Pulmonary (Q_{PA}) 4 78 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

Exploration by catheter showed the right ventricle to be enlarged and the main pulmonary artery to be considerably dilated Since the blood samples from the right auricle and the right ventricle were identical in oxygen content the presence of an interventricular septal defect was ruled out While the catheter was being withdrawn from the pulmonary truncus to the right ventricle, the abrupt change in pressure occurred low within the midsternal shadow (see Fig 30) This indicated that the stenosis was presumably at the infundibular portion of the right ventricle rather than at the pulmonary valve There was a marked degree of systolic hypertension in the right ventricle To explain the fall in pressure between the infundibular region and the left branch of the pulmonary artery the presence of turbulent flow in the dilated post stenotic pulmonary truncus was suggested

GENERAL COMMENTS

The physiological study confirmed the clinical diagnosis and ruled out the presence of other congenital defects The remarkable clinical tolerance of the marked degree of systolic hypertension in the right ventricle should be emphasized

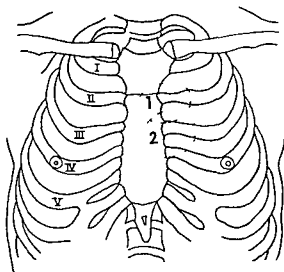
Case 2 ‖ IDIOPATHIC DILATATION OF PULMONARY ARTERY

Clinical data: Age 6 years Weight 22 7 Kg (50 lbs) F

History: Development was normal The child's exercise tolerance was never decreased from normal A murmur was noted shortly after birth

Physical examination: There was a moderately deep funnel depression at the lower sternum No dyspnea or cyanosis was noted on exertion

HEART



Size Increased at base to left

Thrill Moderate systolic along II left inter space

Sounds P only slightly accentuated

Murmurs (See stethogram)

1 Loud, very long, coarse systolic, maximum at II left interspace

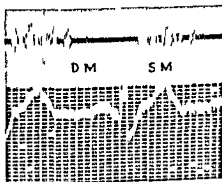
2 Less loud, blowing, decrescendo diastolic, louder in III than II left interspace

Arterial blood pressure 105/70 mm Hg, after exercise, 112/75 mm Hg

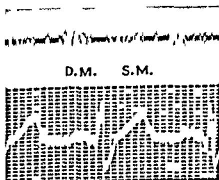
ELECTROCARDIOGRAM

Slight right axis deviation Right bundle branch block, type B (see Lead II, below)

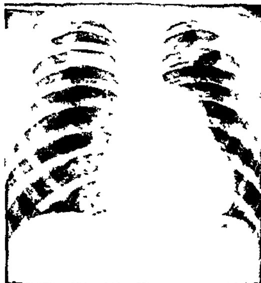
STETHOGRAM



II LEFT INTERSPACE



III LEFT INTERSPACE



P A



L A-O

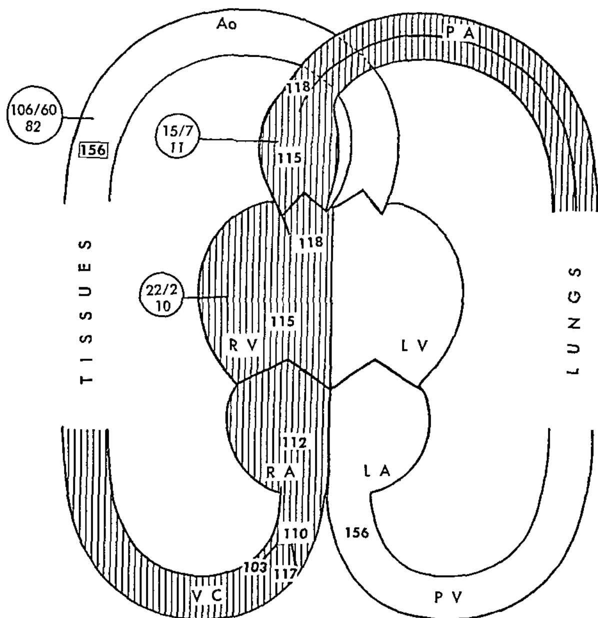


R A-O

There was striking enlargement, though little increase in pulsation, of the main pulmonary artery and its left branch; the other pulmonary vessels were not enlarged or pulsating abnormally. The left ventricle and the auricles did not seem to be enlarged; the right ventricle was slightly enlarged

CLINICAL DIAGNOSIS

Idiopathic dilatation of the pulmonary artery, truncus and left branch, with pulmonic incompetence. This diagnosis, as against that of a patent ductus arteriosus, was based on the following points: (1) the systolic murmur was of equal intensity throughout the phase and lacked intensification at the end of systole, which is frequently noted in cases of a patent ductus; (2) the diastolic murmur was not only entirely different in quality, as it was much more blowing and softer than the systolic, but also differed in location, the point of maximum intensity being below that of the systolic murmur and transmitted well down the left sternal border; (3) the second pulmonic sound was not significantly increased in intensity; (4) the pulse pressure was not wide and the diastolic blood pressure did not drop after exercise; (5) x ray and fluoroscopic findings of a very large but only moderately pulsating main and left pulmonary artery, without enlargement and increased pulsations in the rest of the pulmonary arterial tree.



Arterial blood oxygen capacity — 162 cc/lit
Arterial blood oxygen saturation at rest — 98%
Oxygen consumption at rest = 127 cc/min

ESTIMATED BLOOD FLOW

Systemic (Q_S)	3 10 lit/min
Pulmonary (Q_{PA})	3 10 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

Exploration of the main pulmonary artery and its left branch with the tip of the catheter clearly demonstrated a marked degree of dilatation (see Fig 31) The oxygen content of blood samples from the pulmonary artery and the chambers of the right side of the heart showed no significant difference, indicating that there was no shunt of oxygenated blood at any point Thorough mixing of the venae cavae blood columns was still not complete in the right ventricle The pressure tracings showed a small difference in systolic pressure between the pulmonary artery and the right ventricle This may be chiefly explained by an increase in the expansibility of the pulmonary arteries due to the thinness and dilatation of their walls The role played by turbulent flow is probably less important In the absence of any degree of hypertension in the right ventricle, organic pulmonary stenosis of even mild degree was considered unlikely

GENERAL COMMENTS

The physiological study revealed no evidence of a patent ductus arteriosus and supported the most favored clinical diagnosis of idiopathic dilatation of the pulmonary artery Without angiocardiographic studies it was not possible to decide whether hypoplasia of the ascending aorta was associated with this malformation In the absence of a shunt of highly oxygenated blood above the pulmonary valve the diagnosis of pulmonic incompetence based on the clinical finding of a diastolic murmur could not be confirmed

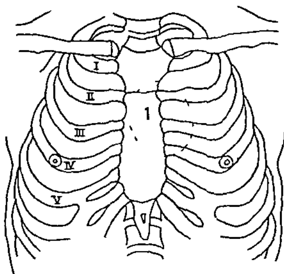
Case 3 ‖ PATENT DUCTUS ARTERIOSUS
MILD HYPERTENSION IN PULMONARY CIRCULATION

Clinical data: Age 3 years Weight 13 6 Kg (30 lbs) F

History: The child had always been very active, without evident dyspnea or cyanosis A murmur was first reported at 1½ years

Physical examination· Her development was normal for her age There was slight pallor

HEART



Size Slightly enlarged

Thrill Systolic, maximum at II and III left inter spaces

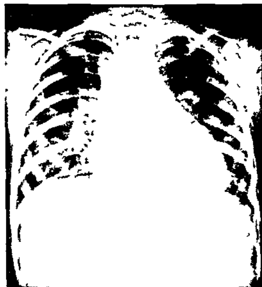
Sounds P_2 masked by murmurs

Murmurs 1 Very loud, continuous machin ery type systolic and diastolic murmurs, maximum at II and III left interspaces, transmitted moderately to right and down ward along left side of sternum No murmurs at apex

Arterial blood pressure 94/48 mm Hg, after exercise, 98/40 mm Hg.

ELECTROCARDIOGRAM

Normal No deviation of electrical axis



P A



L A-O



R A-O

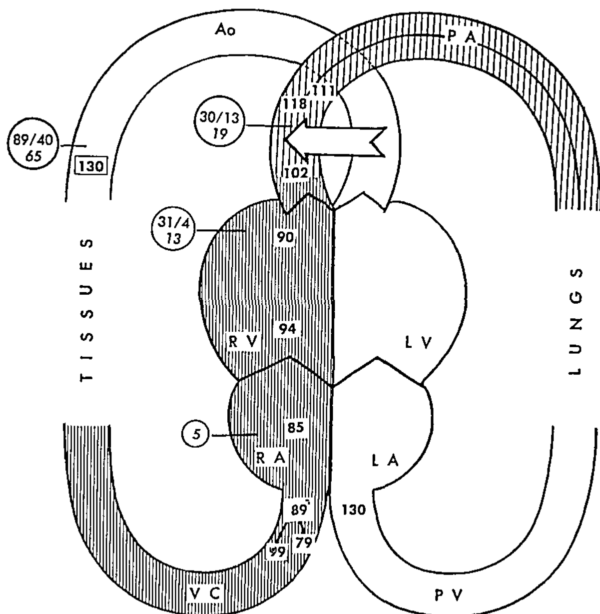


R A-O

The heart showed a slight general increase in size. There was considerable prominence of the left ventricle, especially in the left anterior-oblique view, and of the pulmonary artery segment in the postero-anterior and right anterior-oblique views Slight increase in size of the left auricle was seen in the esophagram. The right auricle was not enlarged. There was an increase in the pulmonary vascular markings in the hilar areas, but no notably expansile pulsations of these vessels were present.

CLINICAL DIAGNOSIS

Patent ductus arteriosus with little enlargement of the heart. This diagnosis was made on the basis of classical types of murmurs, x-ray findings, and wide pulse pressure. In view of the relatively slight increase in pulmonary markings, the amount of shunt through the ductus was thought not to be large.



Arterial blood oxygen capacity — 140 cc/lit
Arterial blood oxygen saturation at rest = 92%
Oxygen consumption (estimated) at rest = 95 cc/min

ESTIMATED BLOOD FLOW		ESTIMATED BLOOD SHUNT	
Systemic (Q_S)	2 38 lit/min	Aortico pulmonary (Y_S)	2 60 lit/min
Pulmonary (Q_{PA})	4 98 lit/min		

COMMENTS ON THE PHYSIOLOGICAL STUDY

The data on oxygen content in the right ventricle and right auricle suggested that there was incomplete mixing in these chambers of the two very different streams of venous blood returning through the superior and inferior venae cavae The marked increase in oxygen content in the truncus and main branches of the pulmonary artery indicated a shunt of oxygenated blood from the aorta Because of its higher oxygen content the blood sample from the right pulmonary artery was probably more representative of blood shunting from the aorta than of truly mixed pulmonary arterial blood The estimated pulmonary blood flow given in the table was based on the oxygen content of the sample in the left pulmonary artery If calculated from the sample from the right pulmonary artery it would be 7 91 lit/min There was only a very slight increase in the mean and diastolic pressures in the pulmonary artery

OPERATIVE FINDINGS

At operation (by Dr Herbert C Maier) a rather short patent ductus about 1 5 cm in diameter was found and ligated with subsequent complete disappearance of the murmurs and thrill

FOLLOW UP

Over the next 9 months there was a decrease in the prominence of the pulmonary segment and the left ventricular shadow and in the accentuation of the pulmonary vascular markings

GENERAL COMMENTS

This is a typical case of patent ductus arteriosus without complications such as pulmonary hypertension and pulmonic incompetence The physiological study confirmed the clinical diagnosis and suggested that the size of the communication and the amount of blood shunting from the aorta were relatively small These findings are consistent with the clinical observations of only a moderate degree of cardiac enlargement and moderate increase of the pulmonary vascular shadows without expansile pulsation It should be noted that with the relatively small increase in the pulmonary flow returning to the left ventricle there was no diastolic murmur heard at the apex

With the pressures in the pulmonary artery almost normal it appears obvious that vascular changes in the pulmonary arterioles and/or hypertension in the somewhat dilated left auricle were minimal

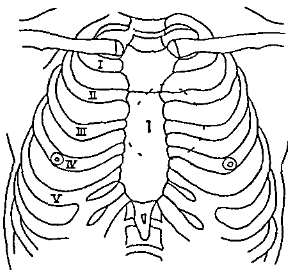
Case 4 — PATENT DUCTUS ARTERIOSUS
MODERATE HYPERTENSION IN PULMONARY CIRCULATION

Clinical data: *Age 3 years Weight 12.7 Kg (28 lbs.) F*

History. *There had been a trace of cyanosis, noted during two attacks of bronchopneumonia* Exercise tolerance was somewhat decreased

Physical examination. The child was slightly underdeveloped There was no clubbing or evidence of cyanosis, but mild dyspnea appeared on fairly active play The breath sounds were increased in the posterior left chest behind the heart

HEART



Size Enlarged

Thrills Coarse, systolic at II and III left inter spaces

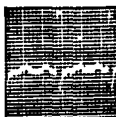
Sounds P_2 increased

Murmurs (See stethogram, page 48)

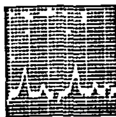
1 Continuous, coarse, "train in tunnel" type, most intense at end of systole and early diastole *Heard widely over upper and* mid precordium, maximum at II and III left interspaces No apical murmurs

Arterial blood pressure 95/15 mm Hg, after exercise, 98/35 mm Hg

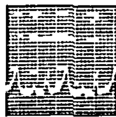
ELECTROCARDIOGRAM



LEAD I



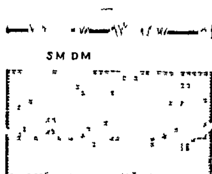
LEAD II



LEAD III

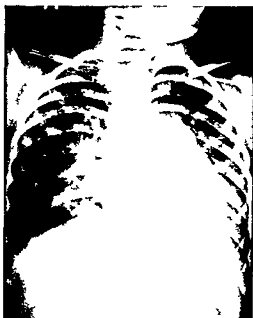
No axis deviation or conduction defect.

STETHOGRAM



II LEFT INTERSPACE

X RAY AND FLUOROSCOPY

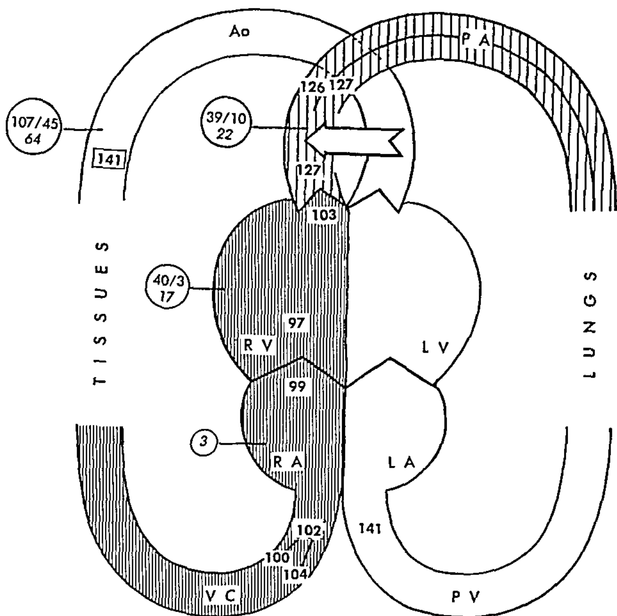


P A

There was general enlargement of the heart, and the apex was low The outflow tract of the right ventricle and the pulmonary artery were enlarged as seen in both the postero anterior and the right anterior oblique view In the left anterior oblique view the left ventricle was quite strikingly enlarged posteriorly There was moderate increase in the size of the right auricle, and the barium filled esophagus showed compression from an enlarged left auricle The lung fields showed considerably increased pulmonary vascular markings, especially in the hilar areas with moderately expansile pulsations

CLINICAL DIAGNOSIS

Patent ductus arteriosus with considerable increase in pulmonary flow and enlargement of the heart. This case exhibited the typical clinical signs (1) classical murmurs (2) wide pulse pressure, (3) drop in diastolic blood pressure after exercise and (4) hilar dance and increased pulmonary vascular markings



Arterial blood oxygen capacity 148 lit/min
Arterial blood oxygen saturation at rest 97%
Oxygen consumption at rest — 95 cc/min

ESTIMATED BLOOD FLOW		ESTIMATED BLOOD SHUNT	
Systemic (Q_s)	2 37 lit/min	Aortico-pulmonary (Y_s)	4 41 lit/min
Pulmonary (Q_{PA})	6 78 lit/min		

COMMENTS ON THE PHYSIOLOGICAL STUDY

The presence of an aortico pulmonary shunt was established from the physiological study, with no evidence of other shunts The blood samples from the main pulmonary artery and both branches were identical in oxygen content, suggesting that the blood was very well mixed The calculation of the pulmonary circulation is therefore reasonably accurate There was no evidence of pulmonic regurgitation of any significance Only a moderate degree of hypertension was present in the pulmonary circulation This was of interest in view of the fairly large calculated blood shunt from the aorta into the pulmonary artery and of a total pulmonary blood flow about three times that of the systemic blood flow

OPERATIVE FINDINGS

At operation (by Dr Charles W Lester) a ductus about 1 cm long and 0 7 cm in diameter was found The pulmonary artery and the outflow tract of the right ventricle were seen to be markedly dilated The ductus was ligated, and the murmurs and thrill disappeared

GENERAL COMMENTS

The physiological study confirmed the clinical diagnosis It is surprising that in this case, where the calculation showed a fairly large pulmonary blood flow and where enlargement of the left auricle and ventricle was noted, there was no diastolic murmur at the apex indicating "relative," non organic mitral stenosis

An increase in pressure in the dilated left auricle was presumed to be the cause of the moderate increase in pressure in the pulmonary artery

It should be emphasized that in the presence of classical clinical signs of a patent ductus arteriosus catheterization of the heart is not necessary as a preoperative diagnostic procedure, unless there is reason to suspect an associated anomaly, particularly pulmonary stenosis

Case 5 | PATENT DUCTUS ARTERIOSUS · INTERVENTRICULAR SEPTAL DEFECT · MARKED PULMONARY HYPERTENSION

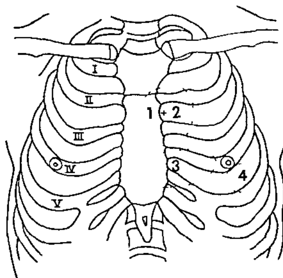
Clinical data Age 8 years Weight 18 4 Kg (40½ lbs) F

History The child had had two attacks of pneumonia and frequent bronchitis She was some what late in walking and had always been less active than other children showing easy fatigue and dyspnea on climbing more than one flight of stairs There was no history suggestive of rheumatic fever

Physical examination Fragile underdeveloped child There was no cyanosis but definite dyspnea appeared with moderate exertion

HEART

Inspection Slight precordial bulge to left of sternum



Size Enlarged downward and out to left.

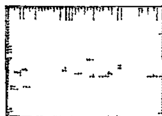
Thrill Moderate systolic maximum at III left interspace

Sounds First at apex increased P markedly accentuated

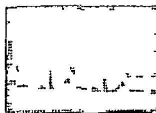
Murmurs
1 Long loud coarse systolic
2 Less coarse early to mid dias tolic both maximum at II left interspace 1 and 2 became con tinuous after exercise
3 Systolic over lower left sternal area transmitted to left higher pitched and less coarse than 1 over pulmonic area
4 Mid diastolic localized at apex different timing and lower pitch than 2 over pulmonic area

Arterial blood pressure 125/76 mm Hg
after exercise 125/48 mm Hg

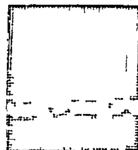
ELECTROCARDIOGRAM



LEAD I

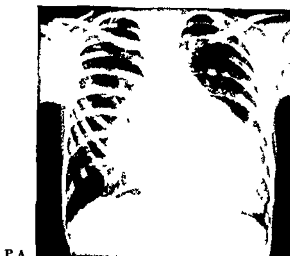


LEAD II



LEAD III

Slight notching of P_1 and P diphasic QRS complexes of increased amplitude in Leads I and III No conduction defect or deviation of the electrical axis



P A



L A-O

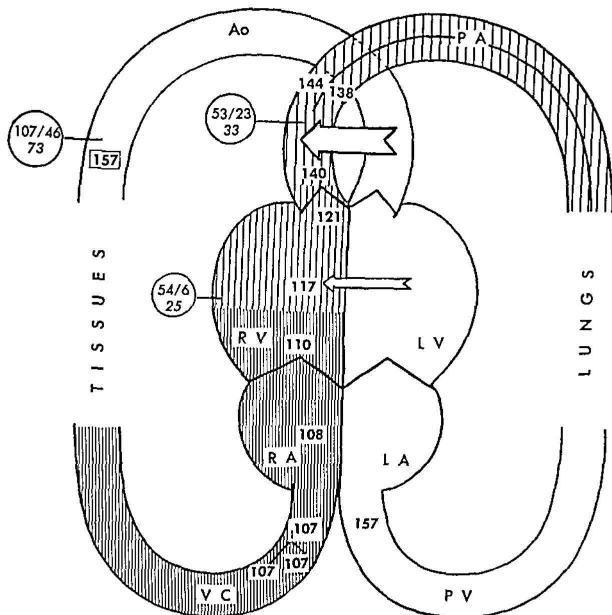


R A-O

X-ray and fluoroscopic examination showed generalized enlargement of the heart. The left ventricle was enlarged outward and down in the postero-anterior view and markedly overrode the spine in the left anterior-oblique view. Enlargement of the right ventricle was observed in all positions. The pulmonary artery segment was prominent, and there was congestion of the smaller pulmonary vessels as well as a moderate degree of expansile pulsation noted in the vessels in the hilar regions Both auricles were definitely enlarged.

CLINICAL DIAGNOSIS

The clinical diagnosis was (1) a patent ductus arteriosus on the basis of murmurs and thrill over the pulmonic area and blood pressure findings; (2) an *interventricular septal defect* based on the finding of a separate systolic murmur over the lower precordium The presence of the second shunt was also suggested by the striking enlargement of the heart, especially the left ventricle. The separate diastolic murmur at the apex was felt to be due to a "relative" mitral stenosis caused by enlargement of the left ventricle and very large pulmonary return flow.



Arterial blood oxygen capacity 173 cc/lit.
Arterial blood oxygen saturation at rest = 92%
Oxygen consumption at rest 152 cc/min

ESTIMATED BLOOD FLOW		ESTIMATED BLOOD SHUNT	
Systemic (Q_S)	3 10 lit/min	Left to right ventricle (Y_2)	0 90 lit/min
Pulmonary (Q_{PA})	9 50 lit/min	Aorta to pulmonary artery (Y_3)	5 50 lit/min

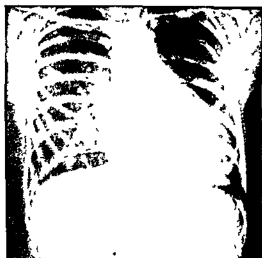
COMMENTS ON THE PHYSIOLOGICAL STUDY

Exploration of the right auricle and ventricle by catheter showed both to be very large A run of auricular tachycardia developed during exploration of the enlarged auricle but subsided after the tip of the catheter was moved

The presence of an arterial shunt from the aorta into the pulmonary artery was demonstrated with a calculated total pulmonary flow of 9 50 lit/min about three times that of the systemic

flow. The difference in oxygen content between the samples taken in the right auricle and the outflow area of the right ventricle could indicate either a shunt of blood from the left to the right ventricle through an interventricular septal defect or pulmonic regurgitation. There was a marked degree of systolic hypertension in the right ventricle and the pulmonary artery.

OPERATIVE FINDINGS AND FOLLOW-UP


PA


LA-O

At operation (by Dr. Herbert C. Maier) the pulmonary artery was found to be considerably enlarged, with a thrill present. The patent ductus arteriosus, measuring approximately 1 cm. in width and 1 cm. in length, was ligated with prompt cessation of the thrill.

After operation the murmurs over the pulmonic area disappeared. As was expected, in view of the interventricular defect, the murmur over the lower precordium persisted with the original intensity and quality. The apical mid-diastolic murmur became very short and faint.

During a 4 months' follow-up period there was a marked gain in weight as well as improvement in cardiac function capacity. The reduction of the vascular markings and the decrease in size of all the cardiac cavities were striking. The child's condition was obviously greatly improved by the elimination of one of the two blood shunts.

GENERAL COMMENTS

This case was of interest on several accounts. (1) In view of the higher oxygenation of the right ventricular blood compared with the right auricular blood in a patient with a patent ductus arteriosus, the presence of a second congenital defect (namely, an interventricular defect) rather than of pulmonic regurgitation, was postulated on the basis of a separate systolic murmur over the lower precordium. For comparison see Case 7, where the diagnosis of pulmonic regurgitation was made on the basis of similar findings in the oxygen content of the blood but different physical signs. The persistence of the systolic murmur over the lower precordium after ligation of the patent ductus in the present case confirmed the original interpretation. (2) The marked decrease in intensity of the mid-diastolic murmur at the apex suggests that after reduction in the pulmonary blood flow, following ligation of the patent ductus, the degree of "relative" mitral stenosis was reduced. (3) The ligation of the patent ductus was followed by decided improvement in the child's general status as well as in her cardiac functional capacity, even in the presence of an interventricular shunt.

The marked elevation of pressures in the pulmonary artery was probably related both to pulmonary vascular changes and to hypertension in the dilated left auricle.

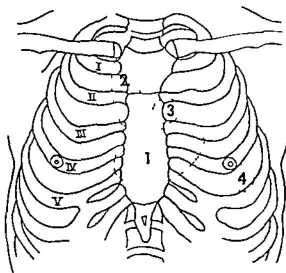
Case 6 — PATENT DUCTUS ARTERIOSUS / HIGH INTERVENTRICULAR SEPTAL DEFECT / FIBROSIS OF AORTIC VALVES

Clinical data: Age 4 years Weight 10 Kg (22 lbs) F

History: The mother had German measles during the sixth week of pregnancy The child had had annual attacks of bronchopneumonia during which only slight cyanosis was present There was moderate dyspnea on exertion

Physical examination: There was marked underdevelopment, with muscular hypotonia, bilateral cataracts, and pigeon breast with deep indentations occurring bilaterally at the level of the diaphragm

HEART



Arterial blood pressure 73/35 mm Hg

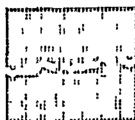
Size Enlarged to left and downward

Thrills 1 Fine systolic at IV left interspace
2 Coarser systolic at suprasternal notch and into neck

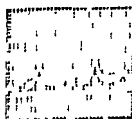
Sounds First at apex very loud A_2 present, equal to P_2

Murmurs 1 Very loud, harsh systolic over lower precordium, maximum at IV left interspace
2 Coarser systolic, maximum at II right interspace, *transmitted to* neck and axilla
3 Very faint, short diastolic at II left interspace
4 Short mid diastolic at apex when supine

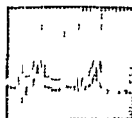
ELECTROCARDIOGRAM



LEAD I

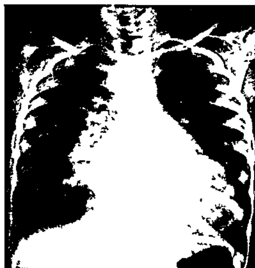


LEAD II

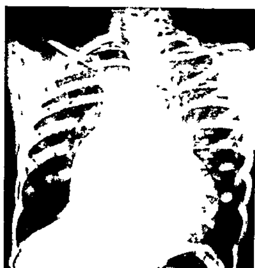


LEAD III

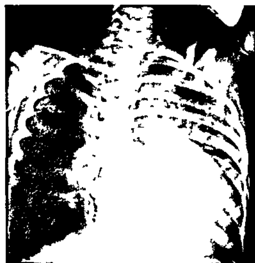
Normal conduction, high T waves and deep Q waves in Leads II and III No significant axis deviation



P-A



L A-O



R A-O

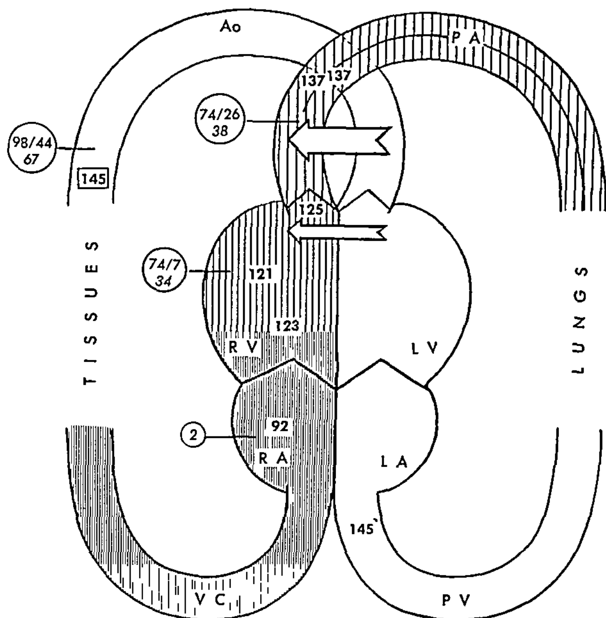


RT. LAT.

There was marked enlargement of the left ventricle in the postero-anterior view and posteriorly in the left anterior-oblique view. The aortic shadow appeared small. The right ventricle and the pulmonary arc were enlarged; increased and collapsing pulsations of the pulmonary vessels were seen. The region of the right auricle was prominent. Note the displacement by chest deformity in the right lateral view.

CLINICAL DIAGNOSIS

Multiple cardiac anomalies: (1) an *interventricular septal defect* as the cause of the systolic murmur over the lower precordium, (2) *subaortic stenosis* causing the systolic murmur described over the right upper precordium, and (3) a *possible patent ductus arteriosus* causing the diastolic murmur at the second left interspace and the wide pulse pressure and adding to the expansile pulsations in the pulmonary vascular shadows. The enlarged size of the right ventricle and the pulmonary vessels was considered due to the increased volume flow resulting from left-to-right shunts through the interventricular septal defect and possibly through a patent ductus. The marked increase in blood return from the lungs would account in part for the enlargement of the left auricle and ventricle, and explain the mid-diastolic murmur at the apex on the basis of a "relative" mitral stenosis. In such a case, with shunting of a large proportion of the circulation into the right heart, the small volume to the systemic circulation would account for the poor general physical development, the low arterial blood pressure, and the small aortic shadow.



Arterial blood oxygen capacity = 153 cc/lit.
Arterial blood oxygen saturation at rest — 96%
Oxygen consumption at rest = 108 cc/min

ESTIMATED BLOOD FLOW		ESTIMATED BLOOD SHUNT	
Systemic (Q_s)	2 04 lit/min	Left ventricle to right ventricle (Y_2)	2 86 lit/min
Pulmonary (Q_{PA})	13 50 lit/min	Aorta to pulmonary artery (Y_3)	8 60 lit/min.

COMMENTS ON THE PHYSIOLOGICAL STUDY

The study demonstrated a large blood shunt from the aorta into the pulmonary artery However, it was considered that the pulmonary flow as calculated might be too high, the blood samples from both branches of the pulmonary artery being more representative of the stream flowing from the aorta to the pulmonary artery than of a complete mixture of blood flowing from the right ventricle and the aorta A marked degree of hypertension was present in the pulmonary circuit. In addition

the marked increase in oxygen content of the right ventricle blood as compared with the right auricle blood was assumed to be due to a fairly large volume of blood shunted from the left to the right ventricle through an interventricular defect. Pressure tracings taken in the brachial artery failed to reveal the plateau like type of curve characteristic of stenosis in the aortic area

OPERATIVE AND AUTOPSY FINDINGS

At operation (by Dr Charles W Lester) the base of the heart was found to be lower than normal, with the apex rotated up in such a way that the arch of the aorta lay almost behind the heart There was a palpable thrill in the aorta, the subclavian artery, and the mediastinum over the ductus During exploration in the region of the ductus the heart stopped suddenly, and attempts at resuscitation failed

At autopsy (courtesy of Dr W W Brandes, Roosevelt Hospital) the chief findings were (1) a high interventricular septal defect 13 mm in diameter, (2) a patent ductus arteriosus, 5 mm in diameter, 13 mm long (3) marked hypertrophy of the left ventricle, moderate hypertrophy of the right ventricle, (4) thickening of the aortic valve cusps, (5) considerable narrowing of the lumina of the pulmonary precapillaries



VIEW OF THE OPEN LEFT VENTRICLE SHOWING THE HYPERTROPHIED MUSCLE AND THE SEPTAL DEFECT IMMEDIATELY UNDER THE FIBROSED AORTIC VALVES

GENERAL COMMENTS

The physiological study in this case demonstrated the presence of a patent ductus arteriosus which had been uncertain on clinical grounds This was confirmed at operation and autopsy The diagnosis of an interventricular septal defect was made both on clinical grounds and from catheterization studies, and was also confirmed at autopsy The fibrosis of the cusps of the aortic valve found at autopsy may explain the site of maximum intensity and direction of transmission of the thrill and systolic murmur at the base, although it is probable that the patent ductus arteriosus contributed to these signs Pulmonary vascular changes were indicated by marked elevation of pressures in the pulmonary artery

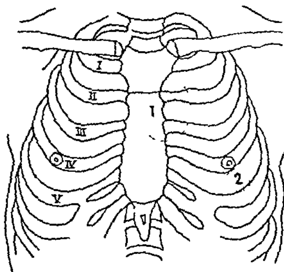
Case 7 ‖ **PATENT DUCTUS ARTERIOSUS AND MARKED PULMONARY HYPERTENSION (STUDIED BEFORE AND AFTER LIGATION)**

Clinical data: Age 3 years Weight 11 8 Kg (26 lbs) F

History: The child had had severe bronchopneumonia at 2½ years, with cyanosis and moderate congestive failure While under continued observation she gradually developed a diastolic murmur over the pulmonic area, a mid diastolic murmur at the apex enlargement of the heart, and increase in pulmonary vascular markings

Physical examination Development was normal Tachycardia and moderate dyspnea were present with exercise

HEART



Size Enlarged to left.

Thrill Slight systolic at II left interspace

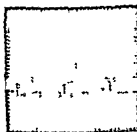
Sounds P_2 increased

Murmurs 1 Rough continuous, machinery type over pulmonic area

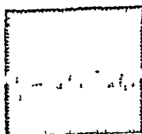
2 Short mid diastolic at apex best heard when supine in left lateral position

Arterial blood pressure 108/60 mm Hg, after exercise, 112/50 mm Hg

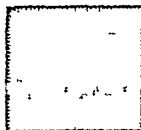
ELECTROCARDIOGRAM



LEAD I



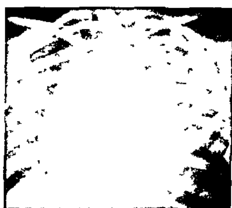
LEAD II



LEAD III

Normal conduction time No axis deviation





P A L A-O

Enlargement of right ventricle and pulmonary arc. Pulmonary vascular markings increased, with hilar dance. Left ventricle increased in size, especially posteriorly. Right auricle moderately enlarged. Left auricle slightly compressed the barium-filled esophagus.

CLINICAL DIAGNOSIS

Patent ductus arteriosus with enlargement of heart. Diastolic murmur at apex interpreted as due to "relative" mitral stenosis caused by large pulmonary flow returning to enlarged left heart.

OPERATIVE FINDINGS

At *operation* (by Dr. Charles W. Lester) *a ductus measuring approximately 1 cm. in length and* 1 cm. in width was found. On ligation of this vessel the thrill previously palpable along the pulmonary artery ceased and the heart rate decreased.

FOLLOW-UP

Five weeks post-operatively: Complete disappearance of murmurs over pulmonic and apical areas. Fluoroscopy and x-rays showed some decrease in the size of the pulmonary artery, which, however, was still quite prominent. Right ventricle only slightly decreased.

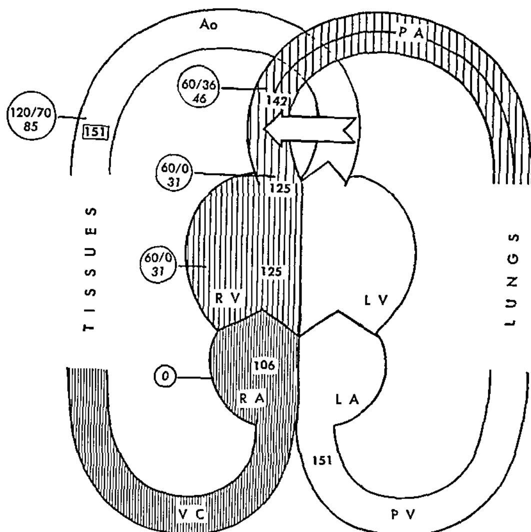
Five months post-operatively: Interval course very satisfactory, with 5 pounds gain in weight. Pulse and respiratory rate normal; returned to base level quickly after vigorous exercise. P_2 no longer accentuated. No murmurs. Blood pressure: 96/60 mm. Hg; after exercise, 100/60 mm. Hg





P A L A-O

X-rays showed a further decrease in the size of the heart and pulmonary artery and normal pulmonary vascular shadows.



Arterial blood oxygen capacity — 162 cc/lit
Arterial blood oxygen saturation at rest = 95%
Oxygen consumption at rest = 92 cc/min

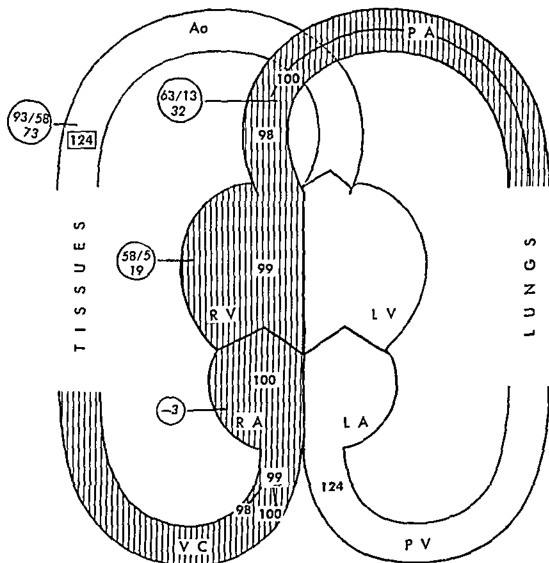
ESTIMATED BLOOD FLOW		ESTIMATED BLOOD SHUNT	
Systemic (Q_s)	2 01 lit/min	Aortico pulmonary (Y_s)	8 16 lit/min
Pulmonary (Q_{PA})	10 20 lit/min	Pulmonic regurgitation (R)	2 28 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

Before operation The oxygen content in the blood samples demonstrated the presence of an arterial shunt into the pulmonary artery The calculated volume of blood shunt was very large and the total pulmonary flow was five times that of the systemic flow There was marked hyper tension, both systolic and diastolic in the pulmonary artery The difference in oxygen content between the right auricle and the right ventricle indicated a flow of highly oxygenated blood into the right ventricle The interpretation could be either an associated interventricular septal

defect or pulmonic incompetence with regurgitation of highly oxygenated pulmonary arterial blood through a dilated pulmonic ring. In the absence of a *separate systolic murmur below the* pulmonic area the diagnosis of pulmonic incompetence was postulated.

POST OPERATIVE CATHETERIZATION—5 WEEKS



Arterial blood oxygen capacity = 133 cc/lit
Arterial blood oxygen saturation at rest = 91%
Oxygen consumption at rest = 92 cc/min

ESTIMATED BLOOD FLOW

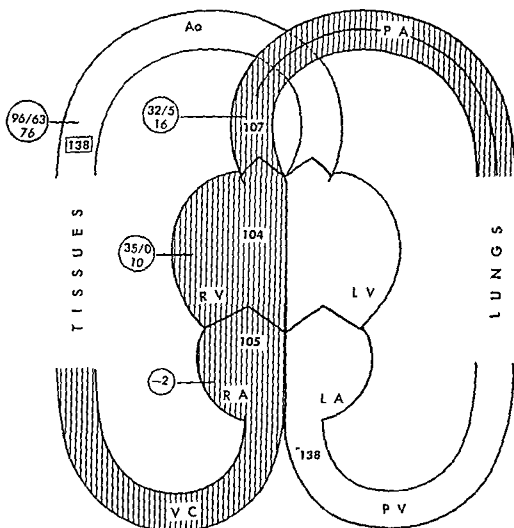
Systemic (Q_s)	3 83 lit/min
Pulmonary (Q_{PA})	3 83 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

Five weeks after operation. There was no longer any evidence of shunt into either the pulmonary artery or the right ventricle. This confirmed the earlier impression that the evidence of

a shunt into the right ventricle found at pre operative catheterization had been due to pulmonic incompetence The systolic pressure of the pulmonary artery was still markedly elevated, while the diastolic pressure had reached a normal level The pulse pressure was therefore very large

POST OPERATIVE CATHETERIZATION—5 MONTHS



Arterial blood oxygen capacity 144 cc/lit
Arterial blood oxygen saturation at rest = 98%
Oxygen consumption at rest 99 cc/min

ESTIMATED BLOOD FLOW

Systemic (Q_s) 3 00 lit/min
Pulmonary (Q_{PA}) 3 00 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

Five months after operation: The pressure in the right ventricle and the pulmonary artery had decreased strikingly and had almost returned to normal

Clinical signs of a patent ductus arteriosus may be less clear cut in infants and young children than in older persons This case presented an opportunity of observing over the period of a year the progressive appearance of the diastolic element and the machinery type murmur as well as a systolic thrill over the pulmonic area During this period there was also clinical evidence suggestive of cardiac insufficiency and of an increasing volume of blood flow through the ductus This was indicated by x ray findings of both relative and absolute increase in the size of the heart and pulmonary artery, and by increasing vascular markings in the lung fields with hilar pulsations of a more collapsing type The development of a mid diastolic murmur limited to the apex in a child who showed no trace of rheumatic infection was interpreted as an added sign of increased return flow from the lungs to the left side of the heart with dilatation of both chambers and swirling of the blood as it passed through an organically normal mitral valve into the enlarged left ventricle

The physiological studies in this case were of interest in showing a very large increase in pulmonary blood flow prior to ligation of the patent ductus arteriosus In this respect the interpretation of the mid diastolic murmur at the apex as being due to increased return flow to the left heart was probably correct as it disappeared after ligation of the ductus when the pulmonary flow was proved to have returned to an almost normal figure

It must be emphasized that in cases of arterial shunt into the pulmonary artery a higher oxygen content in the right ventricle than in the right auricle may be interpreted as the result either of an associated ventricular septal defect or of pulmonic incompetence The latter diagnosis is suggested if there is evidence of considerable increase in pulmonary blood flow with considerable *dilatation of the pulmonary artery The difficulty of separating the systolic murmur of a high* ventricular septal defect from the systolic component of a concomitant patent ductus arteriosus sometimes makes an exact interpretation of blood gas analysis impossible

The presence of a high systolic pressure in the pulmonary artery 5 weeks after ligation of the patent ductus arteriosus at the same level as pre operatively can only be explained by the presence of pulmonary vascular changes due to the very large pulmonary blood flow or to the persistence of left auricular dilatation with hypertension The return to a normal blood pressure in the pulmonary artery after 5 months is evidence that the vascular changes or the left auricular dilatation or both were slowly reversible Furthermore the large pulse pressure observed in the pulmonary artery at the time of the second study was presumptive evidence that pulmonic incompetence was still present It is to be noted that this characteristic finding of valvular incompetence became apparent only after the patent ductus had been ligated and the communication with the aorta during diastole had been eliminated The very large pulse pressure could be alternately *explained by the continuance of the high pulmonary vascular resistance*

Case 8 | INTERAURICULAR SEPTAL DEFECT
MILD PULMONARY STENOSIS
SUBACUTE BACTERIAL ENDOCARDITIS

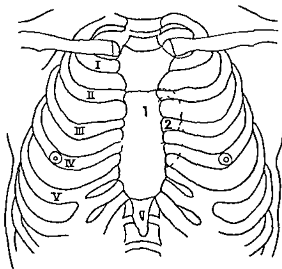
Clinical data Age 5 years Weight 13 6 Kg (30 lbs) F

History The child's physical development had been slow Easy fatigability had always been noted

Present illness Progressive weight loss pallor and fever for 5 weeks Condition critical on admission marked pallor dyspnea and orthopnea present, tachycardia, gallop rhythm heart markedly dilated liver and spleen greatly enlarged Many carious teeth Positive blood cultures for Streptococcus viridans (600 colonies per cc.) confirmed the diagnosis of subacute bacterial endocarditis Congestive failure gradually decreased on digitalis and repeated small transfusions Clinical response to penicillin for 4 weeks was poor although the organism was sensitive in vitro It was therefore most important to decide whether a patent ductus arteriosus was present and if so to ligate it

Physical examination No clubbing or cyanosis Clinical signs of mild failure still present

HEART



Size Enlarged to right and left

Thrill Slight systolic over pulmonic area

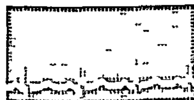
Sounds P slightly increased

Murmurs 1 Long loud harsh systolic at II and III left interspaces

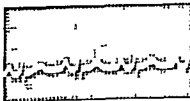
2 Soft blowing diastolic at II III and IV left interspaces

Arterial blood pressure 85/55 mm Hg

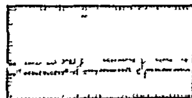
ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

P waves prominent in Lead II No deviation of electrical axis



ON ADMISSION



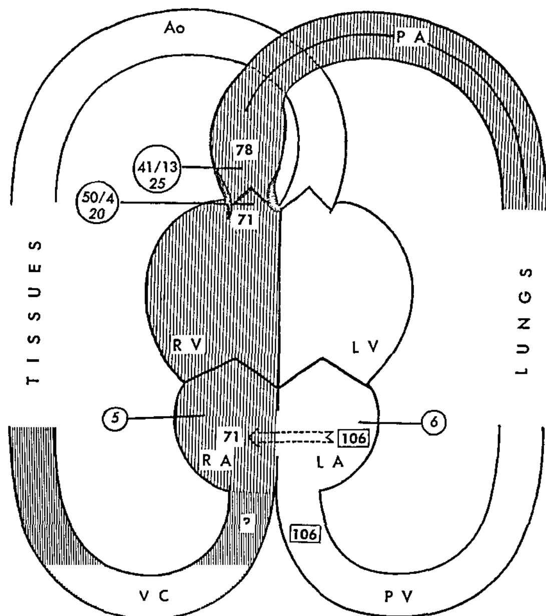


FOUR WEEKS LATER, AT TIME OF FIRST CATHETERIZATION

Time of first catheterization: Generalized enlargement of heart Right auricle and ventricle increased in postero-anterior and left anterior-oblique views; pulmonary artery and branches very prominent with active collapsing type of pulsations noted. Left ventricle enlarged in postero anterior and right anterior-oblique views Left auricle compressed the barium-filled esophagus

CLINICAL DIAGNOSIS

The diagnosis of an *interauricular septal defect with pulmonic regurgitation* was more favored than that of a patent ductus arteriosus, on the basis of the following: (1) the degree of cardiac enlargement, especially of the right auricle, which persisted when other signs of congestive failure were receding; (2) the normal pulse pressure; and (3) particularly the soft, blowing character of the diastolic murmur transmitted down the left sternal border from the pulmonary area The possibility that the clinical signs could be caused either by an interauricular septal defect with pulmonic incompetence or by a patent ductus arteriosus, especially in the presence of moderate congestive failure, was fully recognized and stressed the need for establishing the diagnosis more accurately.



Arterial blood oxygen capacity = 112 cc/lit
Arterial blood oxygen saturation at rest = 96%
Oxygen consumption at rest = 102 cc/min

ESTIMATED BLOOD FLOW

Pulmonary (Q_{PA}) 3 00 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

First catheterization: The presence of an interauricular septal defect was established by passing the tip of the catheter from the right auricle through the left auricle into a left pulmonary vein As blood samples from the superior and inferior venae cavae were not taken, the degree of

shunt from the left auricle to the right auricle could not be estimated nor the systemic blood flow calculated A sample of blood taken from the brachial artery while the child was crying showed an oxygen saturation of 89 per cent, a significant reduction when compared to the oxygen saturation in the left auricular sample obtained while the child was quiet This would indicate a mild degree of right-to-left shunt during physical exertion.

The pressure recordings in the right ventricle indicated a significant degree of hypertension The systolic blood pressure in the pulmonary artery was elevated but somewhat lower than that in the right ventricle.

The diagnosis from the physiological study was an interauricular septal defect with probably a mild degree of pulmonary stenosis. The high pressure in the pulmonary artery was considered related to increased vascular resistance in the pulmonary circuit due to congestive failure

INTERVAL COURSE

Up to time of second catheterization: *Penicillin was increased to 1 million units daily* for 6 weeks with sterilization of the blood and continued clinical improvement up to the time of the second catheterization, done 4 months later.

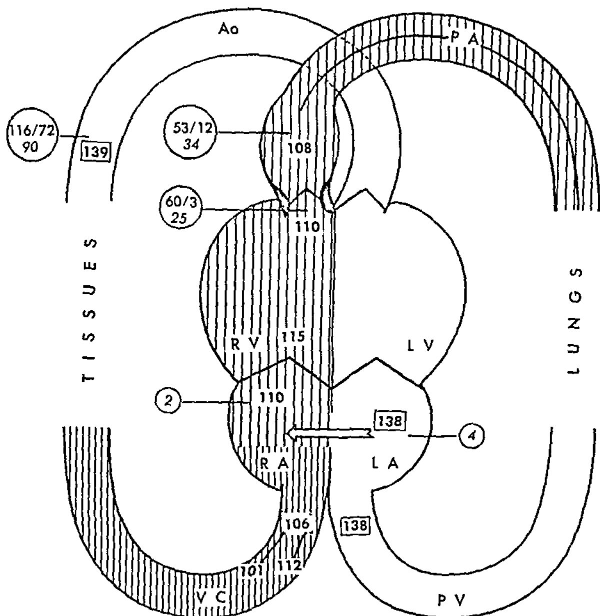
At time of second catheterization:

Clinical data: No signs of congestive failure or active infection Decrease in size of heart Thrill over pulmonic area more pronounced Systolic murmur louder; diastolic murmur shorter and fainter.



X-ray and fluoroscopy: General decrease in size of heart, less congestion in lung fields Collapsing type of pulsations of the main and hilar pulmonary vessels more striking

SECOND CATHETERIZATION



Arterial blood oxygen capacity = 140 cc/lit.
Arterial blood oxygen saturation at rest = 98%
Oxygen consumption at rest = 102 cc/min

ESTIMATED BLOOD FLOW		ESTIMATED BLOOD SHUNT	
Systemic (Q_S)	3 09 lit/min	Left to right auricle (Y_1)	0 31 lit/min
Pulmonary (Q_{PA})	3 40 lit/min		

COMMENTS ON THE PHYSIOLOGICAL STUDY

Second catheterization: The tip of the catheter was again directed into the left auricle and left pulmonary vein The sampling of blood in the left auricle, right auricle, superior and inferior

venae cavae made possible the calculation of systemic blood flow and volume of blood shunted from the left to the right auricle. This shunt was shown to be very small.

A difference in systolic pressure between the pulmonary artery and the right ventricle was again demonstrated on repeated tracings, and the previous finding of pulmonary stenosis of a mild degree was therefore confirmed. There was still a marked degree of hypertension in the pulmonary circuit after clinical signs of failure had cleared. This now strongly suggested that the *cause of the increased* pulmonary resistance was due to permanent changes in the pulmonary vascular tree.

When compared with that of the previous study, the pulmonary blood flow showed an increase. Also, the mean right auricular blood pressure was significantly lower, which was to be expected as the clinical signs of decompensation were no longer present.

GENERAL COMMENTS

By accurately establishing the diagnosis of an interauricular septal defect and eliminating that of a patent ductus, the use of cardiac catheterization prevented an unnecessary exploratory thoracotomy. In view of the extreme rarity of an interauricular septal defect as the site of subacute bacterial endocarditis, the finding of pulmonary stenosis on separate occasions, even though in very mild degree, threw significant light on this case, since a stenotic area is a more likely site of infection. With no shunt of highly oxygenated blood beyond the pulmonary valve, it was impossible to confirm the diagnosis of pulmonic incompetence from the data obtained by catheterization. This diagnosis therefore had to be based entirely on the finding of a characteristic murmur.

The persistence of a high systolic pressure in the pulmonary artery several months after recovery from cardiac failure can be adequately explained only on the basis of pulmonary vascular changes of a permanent nature. In this instance the presumed changes were not related to a large pulmonary blood flow. It is suggested that they may have been secondary to the subacute bacterial endocarditis and the prolonged episode of cardiac failure associated with it. It should be noted that these permanent vascular changes would adequately explain a rise in the systolic pressure in the pulmonary artery as the cardiac output increased following compensation.

In general, the passage of a catheter in the presence of subacute bacterial endocarditis is not recommended. In this case the danger of dislodging a fragment of vegetation by the catheter in the course of the diagnostic procedure was considered less of a hazard than subjecting the child to an exploratory thoracotomy.

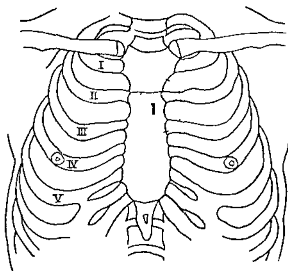
Case 9 INTERAURICULAR SEPTAL DEFECT ANOMALOUS IMPLANTATION OF A PULMONARY VEIN PULMONARY STENOSIS

Clinical data: Age 2½ years Weight 7 5 Kg (16½ lbs) F

History· This child was so markedly retarded that she never held up her head She had had one attack of bronchopneumonia No cyanosis had been observed

Physical examination Mongoloid features and hypotonia Dentition was greatly delayed and defective The palate was cleft

HEART



Size Moderately enlarged to left

Thrill Systolic in I and II left interspaces

Sounds P_2 present although not noticeably accentuated

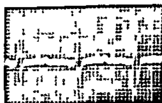
Murmurs 1 Loud rather coarse systolic, maximum at II left interspace, not well transmitted toward left clavicle

Arterial blood pressure 74/50 mm Hg

ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

Right axis deviation No conduction defect

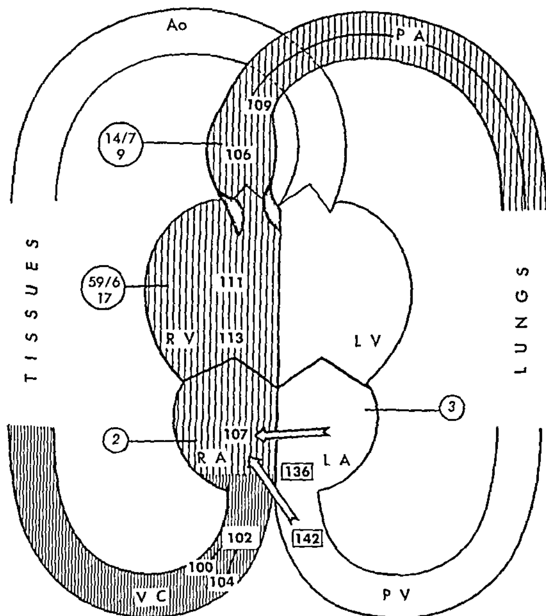

P A


RT. LAT.

The right ventricle was enlarged, and there was fullness of the pulmonary arc but without striking pulsations in the latter area. The right auricle was moderately enlarged in both the postero-anterior and the right anterior-oblique view. No increase in vascular markings or congestion of the lung fields was noted.

CLINICAL DIAGNOSIS

Interauricular septal defect. This diagnosis was based on the location of the murmur and the size of the right auricle, right ventricle, and pulmonary arc as seen in the x-ray.



Arterial blood oxygen capacity 147 cc/lit
Arterial blood oxygen saturation at rest 94%
Oxygen consumption (estimated) at rest — 59 cc/min

ESTIMATED BLOOD FLOW

Systemic (Q_s) 1.17 lit/min
Pulmonary (Q_p) 1.73 lit/min

ESTIMATED BLOOD SHUNT

Pulmonary vein and left auricle to right auricle (Y_1) 0.26 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

The presence of an interauricular septal defect was demonstrated by placement of the catheter in the left auricle and a left pulmonary vein. Direct implantation of a right pulmonary vein in the right auricle was also demonstrated (see Figs. 8 and 11).

The amount of blood shunting from the left to the right side was small, and the calculated pulmonary blood flow was therefore not much increased. The differences in the oxygen content of the various samples from the right cardiac chambers and pulmonary artery were slightly above the upper limit of normal variation. This suggests the persistence of streamline flow as far as the pulmonary artery. In this case the presence of an interauricular septal defect could not have been diagnosed by comparing the oxygen content in both venae cavae and in the right auricle, as there was almost no difference.

Continuous pressure recordings demonstrated stenosis in the infundibular region of the right ventricle and marked elevation of blood pressure in the right ventricle.

GENERAL COMMENTS

Catheterization studies, in addition to confirming the clinical diagnosis of an interauricular septal defect, demonstrated the anomalous implantation of a pulmonary vein and the presence of infundibular pulmonary stenosis which had not been suspected clinically. In retrospect it seems that pulmonary stenosis might have been suggested by the fact that there was no increase in the pulsations of the pulmonary arc and in the vascular markings of the lungs. Post-stenotic dilatation might explain the enlarged pulmonary artery. Furthermore, the demonstration of a fairly small volume of blood shunting into the right auricle and of only a relatively small increase in pulmonary blood flow might well account for the absence of pulsation and of pulmonary congestion. It should be noted that the small shunt into the right auricle was associated with but moderate enlargement of this chamber. In this case of interauricular septal defect with small pulmonary blood flow the finding of pulmonary stenosis explains the degree of enlargement of the right ventricle.

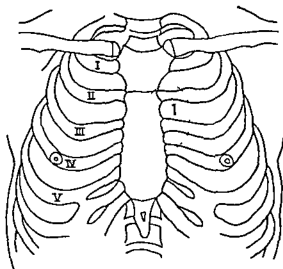
Case 10

IMPLANTATION OF PULMONARY VEINS IN RIGHT AURICL OR INTERAURICULAR SEPTAL DEFECT

Clinical data Age 8 months Weight 4 8 Kg (10½ lbs) F

History This child was markedly retarded (Mongolian idiot) Before her birth her mother had had six other pregnancies the next sibling was eight years older There had been questionable cyanosis on rare occasions during long crying spells

Physical examination There were typical Mongoloid facies and hypotonia The liver edge was felt two fingerbreadths below the costal margin



HEART

Size Enlarged to left and right.

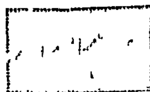
Thrills None felt.

Sounds First sound at apex markedly increased P_2 moderately increased

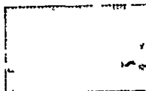
Murmurs 1 Long loud systolic over left precordium maximum at II and III left interspaces

Arterial blood pressure 84/50 mm Hg

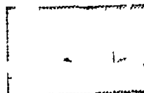
ELECTROCARDIOGRAM



LEAD I

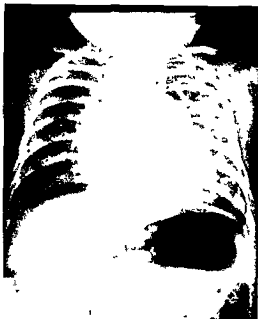


LEAD II



LEAD III

Normal except for peaked P in Lead II



PA



L A-O

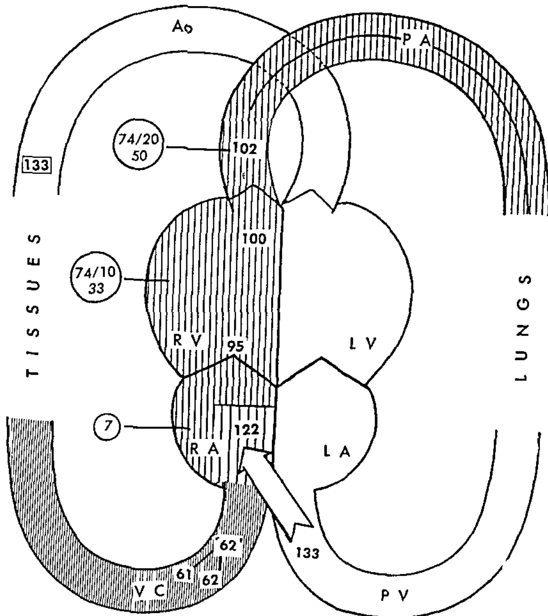


RT. LAT.

There was marked enlargement of the right ventricle, pulmonary arc, and especially of the right auricle. Vascular markings in the lungs were notably increased and striking collapsing pulsations seen on fluoroscopic examination. The left auricle did not compress the barium-filled esophagus.

CLINICAL DIAGNOSIS

Interauricular septal defect. The association of this anomaly with Mongolism has been frequently noted. Because of the large size of the right side of the heart and the striking increase in pulmonary vascular markings with very wide pulsations, it was postulated clinically that there was a large volume of blood shunting into the right heart through the anomaly and an increase in pulmonary blood flow.



Arterial blood oxygen capacity = 137 cc/lit
Arterial blood oxygen saturation at rest = 98%
Oxygen consumption at rest = 41 cc/min

ESTIMATED BLOOD FLOW

Systemic (Q_S)	0 58 lit/min
Pulmonary (Q_{PA})	1 32 lit/min

ESTIMATED BLOOD SHUNT

Pulmonary venous return into right auricle (Y'_1) 0 74 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

Direct exploration with the catheter demonstrated that the size of the right auricle was very greatly increased. The presence of an interauricular septal defect could not be demonstrated, despite repeated attempts to advance the catheter from the right auricle into the left auricle. The presence of a shunt of well-oxygenated blood into the right auricle was predicated upon the large difference in oxygen content between the blood samples from the venae cavae and the right auricle. The oxygen content of samples taken from the right ventricle and the pulmonary artery was lower than that of the sample taken from the right auricle, indicating that the latter must have been withdrawn almost directly from the stream of blood returning through the pulmonary veins The pulmonary flow was over twice the systemic flow. A marked degree of hypertension and wide pulse pressure in the pulmonary circulation were noted (see Fig 28A). A high mean pressure was present in the right auricle, as well as a high end-diastolic pressure in the right ventricle.

GENERAL COMMENTS

An exact diagnosis could not be established in this case beyond the demonstration of a large shunt of well-oxygenated blood into the right auricle. As an interauricular septal defect could not be demonstrated by placement of the tip of the catheter in the left auricle, the alternate diagnosis of anomalous implantation of one or more pulmonary veins in the right auricle was suggested The fact that a blood sample taken from the femoral artery while the child was crying vigorously showed normal oxygen saturation, indicating absence of right-to-left shunt under these circumstances, favors the latter diagnosis If this interpretation is correct, then the systolic murmur heard in the pulmonic area must be entirely accounted for by turbulent flow in the pulmonary artery.

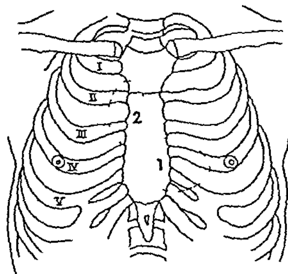
The demonstration by means of pressure recordings of a very wide pulse pressure in the pulmonary artery is of particular interest in correlation with the clinical observation of marked collapsing pulsations seen on fluoroscopic examination. The pulmonary hypertension, in the absence of any evidence of left auricular dilatation, suggests as a mechanism an increased resistance in the pulmonary arterial vessels The association of pulmonary vascular changes and large pulmonary blood flow has often been stressed, although the exact nature of the relationship is not known The degree of pulmonary hypertension has already resulted in changes indicating right-sided failure—namely, an elevated end diastolic pressure in the right ventricle, a notable dilatation of the right auricle, and enlargement of the liver. An important feature of abnormal communication between the right auricle and the left auricle or the pulmonary veins is that continued right-sided failure with the concomitant increase in right auricular pressure will eventually cause pulmonary venous hypertension and congestion.

Case 11 SUBAORTIC STENOSIS
POSSIBLE INTERVENTRICULAR SEPTAL DEFECT

Clinical data Age 11 years Weight 37 3 Kg (82 lbs) M

History The child s development was slow Two siblings were known to have congenital cardiac defects

Physical examination *This boy was slender and showed a moderate degree of mental retardation* He was very active physically without giving any evidence of cyanosis dyspnea or fatigue



HEART

Size Not enlarged

Thrill 1 Systolic at IV left interspace
2 Systolic at II right interspace and up into neck.

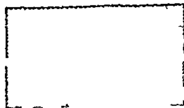
Sounds A present, normal intensity

Murmurs 1 Long harsh systolic at sternal edge in IV and V left interspace transmitted somewhat to left.
2 Rough shorter systolic over aortic area transmitted upward sounded closer to ear than 1

Arterial blood pressure 85/60 mm Hg (arm)
100/65 mm Hg (leg)

ELECTROCARDIOGRAM


LEAD I


LEAD II


LEAD III

Moderate degree of left axis deviation with normal conduction time

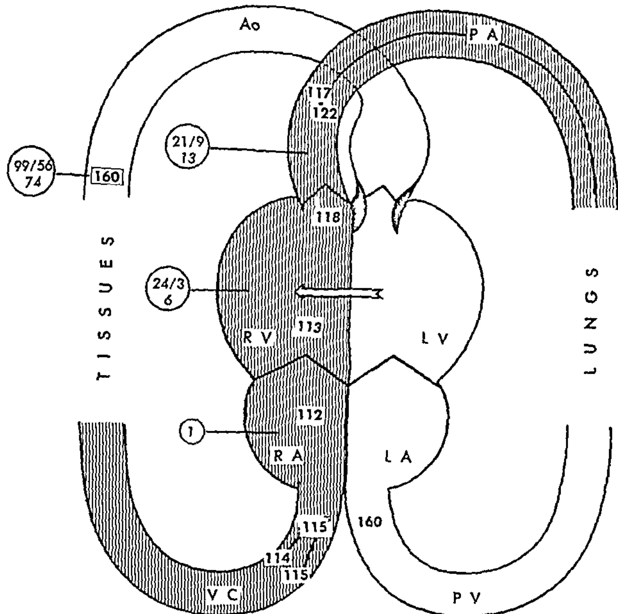


P A L A-O

The heart was not enlarged, but the region of the left ventricle was prominent and there were unusually vigorous contractions The aortic knob was prominent in the postero-anterior view, and the ascending aorta in the left anterior-oblique view.

CLINICAL DIAGNOSIS

The clinical impression, based on the difference in both character and transmission of the systolic murmurs and the presence of two distinct sites of maximum intensity, was that two separate anomalies were involved (1) an *interventricular septal defect,* probably small, as the cause of the murmur at the lower sternal edge; (2) *subaortic stenosis,* suggested by transmission of murmur into the area of the neck, distinct second aortic sound, low blood pressure with narrow pulse pressure, evidence of slight dilatation of the aorta (post-stenotic), and left axis deviation



Arterial blood oxygen capacity		174 cc/lit
Arterial blood oxygen saturation	at rest	94%
Oxygen consumption	at rest	154 cc/min

ESTIMATED BLOOD FLOW		ESTIMATED BLOOD SHUNT	
Systemic (Q_S)	3.21 lit/min	Left to right ventricle (Y)	0.54 lit/min
Pulmonary (Q_{PA})	3.75 lit/min		

COMMENTS ON THE PHYSIOLOGICAL STUDY

The difference in oxygen content between the blood samples taken in the right auricle and the outflow tract of the right ventricle was thought to indicate shunting of a small amount of blood from the left ventricle into the right ventricle The fact that the oxygen content was higher in the two branches of the pulmonary artery than in the right auricle and tricuspid area of the right ventricle would also seem to support this view

The records of the pressures taken from both the femoral and the brachial artery on two separate occasions showed the plateau like curve characteristic of narrowing at the aortic ring or in the aorta (see Fig 32) The blood pressures in the pulmonary artery and right ventricle were normal

GENERAL COMMENTS

The arterial blood pressure tracings support the clinical diagnosis of stenosis in the region of the aortic ring Confirmation of the diagnosis of an interventricular septal defect is less certain from the physiological data as the differences in oxygen content in the various blood samples were just at the limit of significance

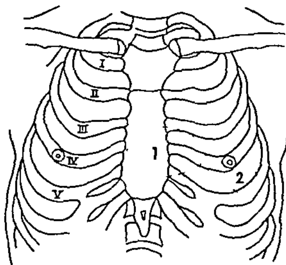
Case 12 ‖ INTERVENTRICULAR SEPTAL DEFECT MILD PULMONARY HYPERTENSION

Clinical data Age 3 years Weight 12 3 Kg (27 lbs) F

History The child s development had been normal but exercise tolerance had always been somewhat decreased "Two murmurs had been present since early infancy Although there was no history suggestive of active rheumatic fever this case had been previously diagnosed elsewhere as rheumatic mitral insufficiency and stenosis

Physical examination There was no clubbing Dyspnea was noted after moderate exercise but no cyanosis was present

HEART



Size Enlarged to left at apex.

Thrill Rather coarse systolic over lower precordium maximum at IV left interspace near sternum

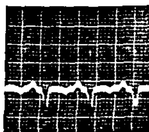
Sounds P accentuated

Murmurs 1 Long loud coarse systolic maximum at IV left interspace transmitted both beyond right side of sternum and to left beyond apex

2 Short mid diastolic at and outside apex when supine increased after exercise

Arterial blood pressure 95/65 mm Hg

ELECTROCARDIOGRAM



LEAD I

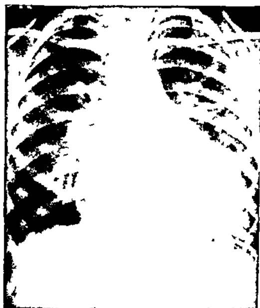


LEAD II



LEAD III

Slight left axis deviation



P A



L A-O

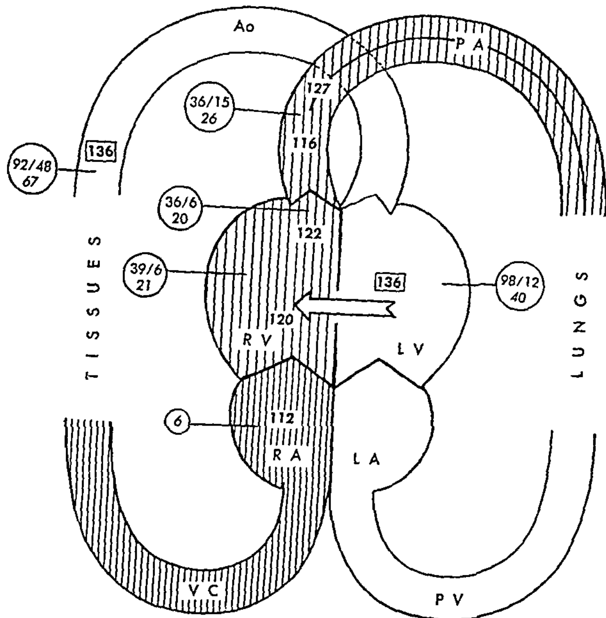


R A-O

There was generalized cardiac enlargement with striking fullness in the region of the pulmonary arc in the postero-anterior view. Both ventricles were enlarged, the left somewhat more than the right in the postero-anterior and oblique views The right auricular curve was considerably accentuated in the postero-anterior view, and there was moderate displacement of the barium-filled esophagus by the left auricle. The aorta appeared small. There was engorgement of the pulmonary vessels, but no striking degree of collapsing pulsation could be seen in the hilar areas.

CLINICAL DIAGNOSIS

Interventricular septal defect This defect was thought to be of large size with considerable shunt into the right ventricle and large flow through the pulmonary circuit The diastolic murmur at the apex was considered to be due to a "relative," or physiological, mitral stenosis dependent on the large return flow from the lungs into the enlarged left auricle and ventricle.



Arterial blood oxygen capacity 145 cc/lit
Arterial blood oxygen saturation at rest = 95%
Oxygen consumption at rest = 86 cc/min

ESTIMATED BLOOD FLOW

Systemic (Q_s) 3 57 lit/min
Pulmonary (Q_{PA}) 6 14 lit/min

ESTIMATED BLOOD SHUNT

Left ventricle to right ventricle (Y) 2 57 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

An interventricular septal defect was demonstrated by (1) directing the tip of the catheter from the right ventricle to the left ventricle and identifying this location by x ray in the oblique position (see Figs 12 and 13), by (2) pressure recording (see Fig 25), and by (3) analysis of blood samples. The level at which the tip of the catheter was passed into the left ventricle is presumptive evidence that the defect was located low in the septum rather than in the membranous part. A mild degree of hypertension in the pulmonary circulation was present. The pulmonary flow was almost twice the amount of the systemic flow. The increase in oxygen content between the blood sample from the right auricle and that from the right ventricle was at most 10 cc/lit. The variation of oxygen content in samples from the main and left pulmonary arteries was most probably due to laminar flow. While the tip of the catheter was in the left ventricle a short run of ventricular tachycardia was observed (for details see page 13)

GENERAL COMMENTS

The physiological study confirmed the presence of an interventricular defect near the apex with a fairly large increase in pulmonary circulation. In this case demonstration of a large pulmonary return flow would account for the mid diastolic murmur at the apex. This case presents a contrast to the type of interventricular septal defect described as maladie de Roger, because of the marked enlargement of the heart and the decrease in functional capacity. This may well be explained by the demonstration of a large shunt which would suggest a larger than average defect in the septum.

The mild degree of hypertension in the pulmonary artery can be related in this instance to the increased end diastolic or filling pressure in the left ventricle and presumably to the increased pressure in the left auricle and pulmonary veins. Whether dynamic adjustments to the shunt contribute to the rise in pressure in the right ventricle and also to the drop in systolic pressure in the left ventricle is in the nature of an interesting speculation.

The following correlation between physical signs and findings during the physiological study is worthy of note—namely that the exact point of maximum intensity of systolic thrill and murmur corresponded to the level checked by fluoroscopy at which pressure recording demonstrated that the tip of the catheter was passing through the defect from one ventricle to the other.

It is to be emphasized that when there is only a small difference in oxygen content between the systemic arterial blood and the mixed venous blood sampled in the right auricle even the addition of a large amount of fully oxygenated blood through a shunt does not produce a marked absolute increase in the oxygen content of the right ventricular blood.

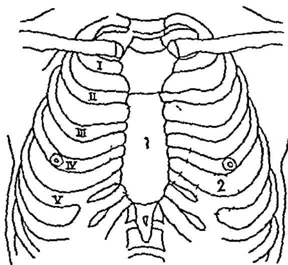
Case 13 INTERVENTRICULAR SEPTAL DEFECT INFUNDIBULAR PULMONARY STENOSIS

Clinical data: Age 10 years Weight 30 Kg (66 lbs) M

History. *The child had always had moderate dyspnea on exertion and occasionally slight cyanosis* There had been nothing in the history suggestive of rheumatic fever

Physical examination. The stature was small Exercise tolerance was decreased

HEART



Size Enlarged to left and right. Moderate precordial bulge

Thrill Systolic, coarse all over precordium maximum at III and IV interspaces

Sounds P slightly louder than A.

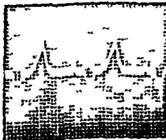
Murmurs 1 Loud harsh systolic, maximum at III and IV interspaces right and left of sternum, widely transmitted

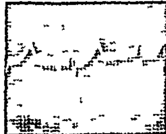
2 Loud mid diastolic at apex.

Arterial blood pressure 90/62 mm Hg

ELECTROCARDIOGRAM


LEAD I


LEAD II


LEAD III

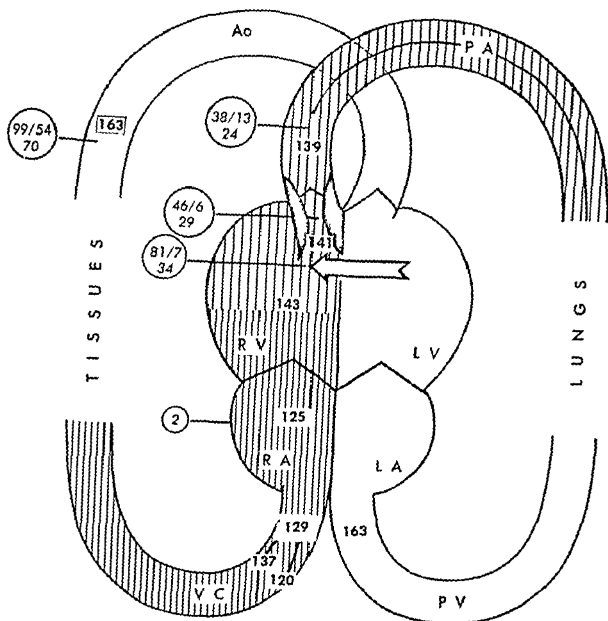
No electrical axis deviation T very high and peaked



There was considerable enlargement of the right ventricle and right auricle in the postero-anterior and left anterior-oblique views. The pulmonary conus was not prominent. The interventricular groove and left ventricle were displaced posteriorly. The left auricle was slightly enlarged. Pulmonary vascular markings were accentuated in the hilar areas, but no expansile pulsations were noted. The aorta was small.

CLINICAL DIAGNOSIS

Interauricular septal defect with congenital mitral stenosis. Lutembacher's syndrome, but without pulmonary artery enlargement. This impression was based on (1) the size of the right auricle, which appeared very large by x-ray, and (2) the presence of an unusually loud diastolic murmur at the apex in the absence of any rheumatic history.



Arterial blood oxygen capacity 166 cc/lit
Arterial blood oxygen saturation at rest 99%
Oxygen consumption at rest = 130 cc/min

ESTIMATED BLOOD FLOW		ESTIMATED BLOOD SHUNT	
Systemic (Q_s)	3 42 lit/min	Left to right ventricle (Y_r)	2 00 lit/min
Pulmonary (Q_{PA})	5 42 lit/min		

COMMENTS ON THE PHYSIOLOGICAL STUDY

Exploration of the right auricle with the tip of the catheter showed this chamber to be only moderately enlarged but displaced to the right by an enormously enlarged right ventricle No abnormal communication between the auricles could be demonstrated The difference in oxygen content between samples withdrawn from the right auricle and the right ventricle indicated the presence of a shunt from the left to the right ventricle The pulmonary blood flow was only moderately increased The pressure recordings taken continuously in the pulmonary artery, the outflow tract and the middle portion of the right ventricle definitely indicated the presence of stenosis in the pulmonary infundibular area

GENERAL COMMENTS

The physiological study eliminated the clinical diagnosis of an interauricular septal defect By exploration with the catheter it was possible to demonstrate visually that the large shadow in the location of the right auricle was due less to intrinsic increase in the size of this chamber than to displacement by a very large right ventricle

In this case of interventricular septal defect complicated with pulmonary stenosis, the marked hypertension in the right ventricle and the striking increase in the size of this chamber may be largely explained by the factor of pulmonary stenosis The apical diastolic murmur was thought to be due to "relative" mitral stenosis in association with a moderate increase in pulmonary blood flow, although the diagnosis of an organic mitral stenosis cannot be entirely discarded The slight systolic hypertension present in the pulmonary artery is probably related to a blood pressure increase in the left auricle, which was demonstrated clinically as somewhat dilated

Case 14 ‖ INTERVENTRICULAR SEPTAL DEFECT MARKED PULMONARY HYPERTENSION

Clinical data Age 12 years Weight 50 Kg. (110 lbs) M

History A murmur was noted in infancy The child s physical activity was restricted up to 6 years of age but afterward he played almost as actively as other children with no fatigue

Physical examination Very well developed boy No clubbing was present No cyanosis or dyspnea were noted after considerable exercise The liver was not enlarged

HEART

Size Slightly enlarged to left at apex.

Thrill Striking rather fine systolic, maximum at III left interspace near sternum radiating down but more upward and out to left.

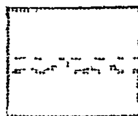
Sounds P markedly accentuated thudding

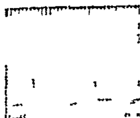
Murmurs 1 Long loud coarse systolic maximum at III left interspace transmitted maximally toward clavicle less loudly to IV inter space

2 Systolic over lower sternal area louder on left than right less in tense and lower in pitch than I over pulmonic area Systolic murmur heard also over left su prascapular area

Arterial blood pressure 102/80 mm Hg no drop in diastolic level after exercise

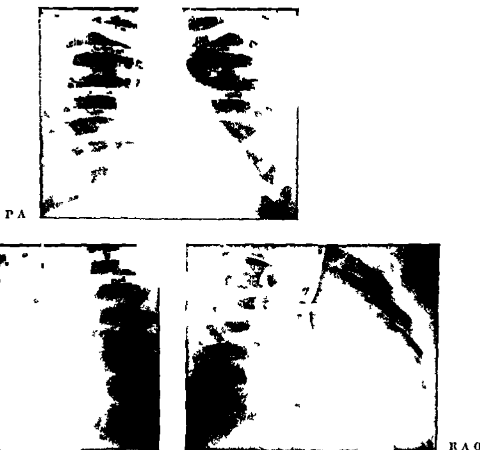
ELECTROCARDIOGRAM


LEAD I


LEAD II


LEAD III

Right axis deviation Normal conduction time



PA

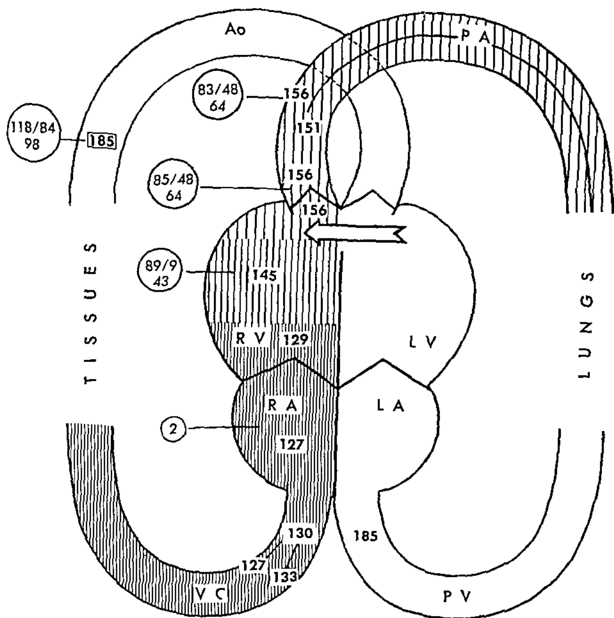
LAO RAO

Left ventricle enlarged downward in postero anterior view and posteriorly in left anterior oblique view Right ventricle moderately enlarged Pulmonary artery enlarged and increased and expansile pulsations seen well out in the smaller branches Right auricle only slightly prominent left auricle not enlarged Aortic knob not visible

CLINICAL DIAGNOSIS

Interventricular septal defect with mild to moderate degree of pulmonary stenosis probably in infundibular area This diagnosis was based on the difference in quality and intensity of the systolic murmurs present over the lower and upper precordium The notable enlargement and expansile pulsations of the pulmonary vessel suggested a large pulmonary blood flow

Other diagnoses discussed were (1) patent ductus arteriosus (2) interauricular septal defect with large pulmonary artery and a concomitant pulmonary stenosis of mild degree The former was considered unlikely in view of (a) the fact that a diastolic murmur had not developed though the child was 12 years old (b) the relatively narrow pulse pressure and the absence of a drop in diastolic pressure after strenuous exercise The latter was ruled out partly because of the position of the murmur heard over the lower precordium but chiefly because the right auricle did not appear significantly enlarged



Arterial blood oxygen capacity = 192 lit/min
Arterial blood oxygen saturation at rest = 97%
Oxygen consumption at rest = 215 cc/min

ESTIMATED BLOOD FLOW		ESTIMATED BLOOD SHUNT	
Systemic (Q_s)	3 70 lit/min	Left to right ventricle (Y .)	3 25 lit/min
Pulmonary (Q_{PA})	6 95 lit/min		

COMMENTS ON THE PHYSIOLOGICAL STUDY

The presence of a blood shunt from the left to the right ventricle was demonstrated by analysis of the oxygen content in blood samples There was marked systolic hypertension in the right ventricle and the pulmonary artery The end diastolic pressure in the right ventricle was found to be markedly elevated Continuous pressure recordings taken while the catheter was moved back and forth several times across the pulmonary valve area did not reveal any evidence of pulmonary stenosis the systolic pressure remaining identical

GENERAL COMMENTS

The physiological study confirmed the clinical diagnosis of an interventricular septal defect but failed to demonstrate any degree of pulmonary stenosis The presence of a patent ductus or an interauricular septal defect was ruled out

One of the striking features in this case is the marked hypertension in the pulmonary circulation with no evidence of pulmonary stenosis (for comparison see Cases 12 and 13) The remarkable tolerance of such a degree of pulmonary hypertension should be stressed It is to be noted that the end diastolic pressure in the right ventricle was shown to be elevated above normal this elevation of filling pressure would appear to be a function of increased flow rather than of beginning decompensation

The high pressure in the pulmonary artery must be explained on the basis of significant pathological changes in the pulmonary arterial system which caused an increased resistance to flow

If it is assumed that blood is ejected directly into the infundibular area and the pulmonary artery through a high interventricular septal defect then favorable dynamic conditions would be present in the pulmonary artery for the creation of a second systolic murmur in this region distinct from the systolic murmur caused by flow through the septal defect

It should be pointed out that the combination of a high ventricular septal defect with enlargement of and high pressures in the pulmonary artery if associated with some degree of overriding of the aorta constitutes the Eisenmenger complex In cases where cyanosis is not present the latter diagnosis is established upon the demonstration of a slight degree of oxygen unsaturation in the arterial blood under conditions of rest and further decrease in saturation during exercise while the patient is breathing pure oxygen

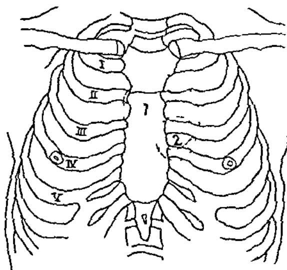
Case 15 ‖ HIGH VENTRICULAR SEPTAL DEFECT WITH PROBABLE AORTIC SEPTAL DEFECT MARKED PULMONARY HYPERTENSION

Clinical data Age 7 years Weight 15 4 Kg (34 lbs) F

History The child s physical development and weight gain were very slow She had had pneumonia four times slight cyanosis was noted then She has always shown considerable fatigue and some dyspnea on moderate exertion

Physical examination Underdeveloped thin fragile looking child No clubbing was present. Exercise tolerance was considerably decreased

HEART



Inspection Precordial bulge mostly on left side deep indentations of anterior chest wall at diaphragm level

Size Enlarged apex at anterior axillary line in VI interspace Increased size to left in pulmonary area

Thrill Slight systolic in II left interspace

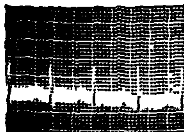
Sounds P *markedly accentuated*

Murmurs 1 Long loud rather harsh systolic over upper precordium maximum at II and III left interspace

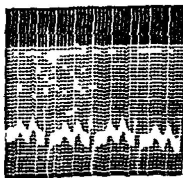
2 Early diastolic maximum at III left interspace, shorter and softer than 1 became continuous with slight exercise

Arterial blood pressure 90/50 mm Hg after exercise 90/30 mm Hg

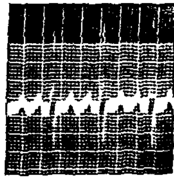
ELECTROCARDIOGRAM



LEAD I



LEAD II

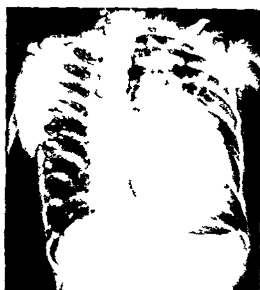


LEAD III

High P waves in Lead II



P A



R A O



L A O

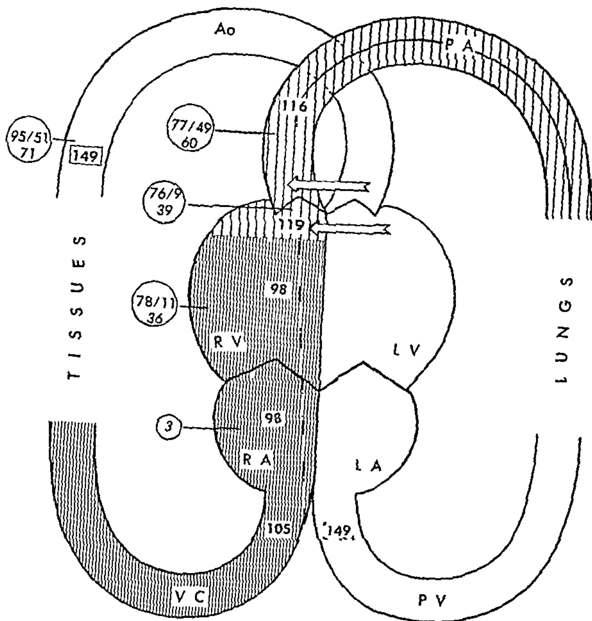


RT LAT

Heart somewhat displaced to left *Depression at lower precordium seen in lateral x ray* The right ventricle showed enlargement in both the postero anterior and left anterior-oblique views, the pulmonary artery was notably enlarged and pulsated very actively The left ventricle was enlarged downward and posteriorly *The right auricle* appeared moderately *enlarged*, *the left auricle* slightly so The secondary and tertiary pulmonary vessels were considerably *enlarged but the* pulsations, though vigorous, were not collapsing in type.

CLINICAL DIAGNOSIS

The diagnosis rested between (1) a *patent ductus arteriosus* with an unusual degree of dilatation of the pulmonary artery and possible pulmonic incompetence and (2) an *interauricular septal defect* with an enlarged pulmonary *artery and pulmonic* incompetence Because of the wide pulse pressure and drop in diastolic pressure after exercise, the first diagnosis was considered the more likely, despite the marked cardiac enlargement.



Arterial blood oxygen capacity 149 cc/lit
Arterial blood oxygen saturation at rest 100%
Oxygen consumption at rest = 103 cc/min

ESTIMATED BLOOD FLOW

Systemic (Q_S)	2 02 lit/min.
Pulmonary (Q_{PA})	3 32 lit/min

ESTIMATED BLOOD SHUNT

Left to right ventricle (Y)
combined (?) with aortico pulmonary } 1 30 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

Exploration of the right auricle showed dilatation of this chamber, but repeated efforts to place the catheter tip in the left auricle failed to reveal a communication between the auricles The results of blood gas analysis were interpreted as indicating the presence of an aortico pulmonary shunt with pulmonic regurgitation The marked degree of hypertension both systolic and diastolic in the pulmonary artery was considered remarkable in view of the small calculated shunt from the aorta to the pulmonary artery

OPERATIVE FINDINGS

On the basis of the physiological findings an exploratory thoracotomy through a postero lateral incision to provide wide exposure was performed (by Dr Herbert C Maier) A very large pulmonary artery was found but no thrill could be felt over the dilated pulmonary artery or over the aorta No patent ductus arteriosus was present The child recovered from the exploration without difficulties

GENERAL COMMENTS

An interauricular septal defect was ruled out by the physiological studies In view of the operative findings the diagnosis of a patent ductus arteriosus can be discarded

The exact nature of the shunt between the left and right ventricles and/or between the aorta and the pulmonary artery has still not been proven To fit the physiological and clinical data there are two possible diagnoses to consider (1) a high interventricular septal defect with pulmonic incompetence and (2) the combination of a high interventricular septal defect and an aortic septal defect The first diagnosis seems less likely than the second in view of (a) practically identical diastolic pressures in the pulmonary and brachial arteries and (b) the fall of the diastolic pressure in the brachial artery following exercise (for comparison see Case 14)

In view of the minimal enlargement of the left auricle, the high pulmonary arterial pressures must be related here to significant pathological changes in the pulmonary arterial bed

It should be mentioned that although the pulmonary blood flow was not great at the time of study the degree of pulmonary hypertension and the presumed vascular changes suggest the *presence of a much larger shunt at an earlier period In general* an increasing resistance in the vascular bed should tend to limit the volume of left to right shunt

Case 16

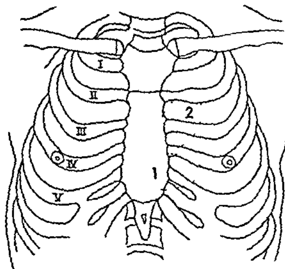
INFUNDIBULAR PULMONARY STENOSIS
INTERVENTRICULAR SEPTAL DEFECT
OVERRIDING OF THE AORTA (MARKED DEGREE)
RIGHT AORTIC ARCH

Clinical data: Age 6 years Weight 26 3 Kg (58 lbs) M

History: The child had been cyanotic since the first month of life His exercise ability was markedly limited, he preferred to move around 'on all fours' and squatted frequently when walking erect The history included two episodes of auricular paroxysmal tachycardia

Physical examination: General development and nutrition were good There was moderate clubbing of nails Cyanosis was present at rest, with slight exertion the cyanosis deepened markedly and the child became quite dyspneic

HEART



Size Not enlarged

Thrill None felt

Sounds P decreased clear

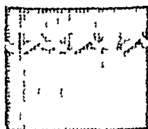
Murmurs 1 Blowing systolic, low over sternum

2 Higher pitched systolic, maximum at III left interspace

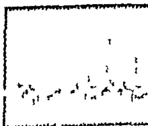
Arterial blood pressure 90/60 mm Hg

Hb 17 0 Gm , R B C 7 01 million

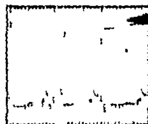
ELECTROCARDIOGRAM



LEAD I



LEAD II



LEAD III

Marked right axis deviation



P A

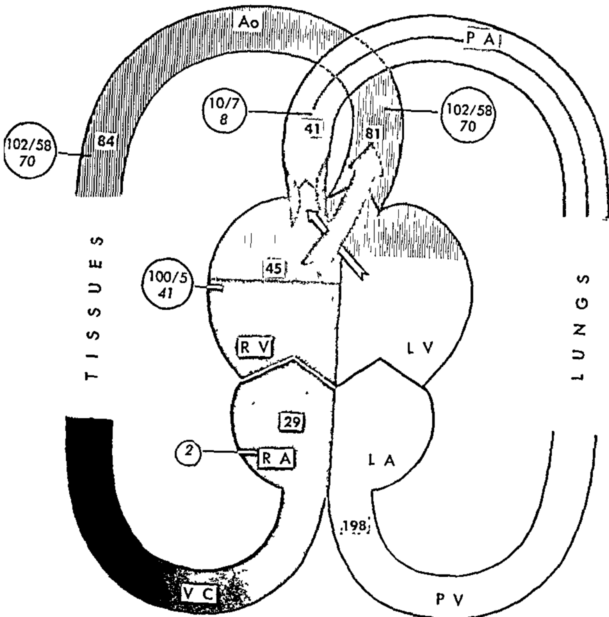


R A O

Examination by x ray and fluoroscopy showed a rounded and raised apex and only a moderate degree of concavity in the region of pulmonary arc with no pulsations visible Fluoroscopy revealed increased clarity of the pulmonary window in the left anterior oblique view The lung fields were strikingly clear The aortic arch indented the barium filled esophagus on the right as poorly illustrated above in the postero anterior view

CLINICAL DIAGNOSIS

The clinical diagnosis was *tetralogy of Fallot with right aortic arch*



Arterial blood oxygen capacity . . = 193 cc/lit.
Arterial blood oxygen saturation at rest = 43%
Oxygen consumption . at rest = 110 cc/min.

ESTIMATED BLOOD FLOW

Systemic (Q_s) . . . . . 2 0 lit/min.
Pulmonary (Q_{PA}) . . greater than 0 70 and smaller than 0 91 lit/min.

ESTIMATED BLOOD SHUNT

Right ventricle to aorta (X) . . . . 1.31 lit/min
Left ventricle to right ventricle (Y'_2) . . . 0 01 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

Overriding of the aorta and a right aortic arch were demonstrated by directing the tip of the catheter from the right ventricle into the aorta and beyond the arch (see Fig 16).

From the analysis of the oxygen content in the blood samples it was calculated that the shunt of blood from the left ventricle to the right ventricle was small and that the shunt from the right ventricle to the aorta was large The latter finding suggested a marked degree of overriding of the aorta.

The pressure tracings showed a marked degree of stenosis in the infundibular area with an extremely low pressure in the pulmonary artery and a very high systolic pressure in the body of the right ventricle The systolic pressures in the right ventricle, aorta, and brachial artery were almost identical The systemic blood flow was somewhat reduced The pulmonary blood flow was at most only half the systemic flow.

OPERATIVE FINDINGS

At operation (by Dr Alfred Blalock) the diagnosis of a right aortic arch was confirmed The left pulmonary artery was fairly large but collapsed easily. Anastomosis was made between the end of the subclavian and the side of the pulmonary artery A marked thrill was then felt in the pulmonary artery.

FOLLOW-UP

The child has improved markedly since operation His physical activity has been greatly increased, and the cyanosis, the dyspnea, and the polycythemia have been relieved

GENERAL COMMENTS

The physiological study added the following information to the diagnosis already made on clinical grounds: (1) that the location of the stenosis was infundibular, (2) that the maximum amount of flow through the pulmonary capillaries (combining the pulmonary artery and the assumed collateral flow) did not exceed 1 liter per minute

Case 17

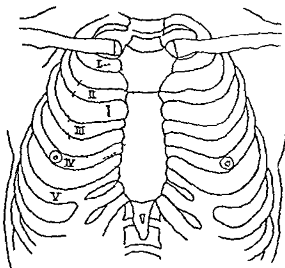
DEXTROCARDIA WITH RIGHT AORTIC ARCH
DOUBLE SUPERIOR VENA CAVA
SINGLE AURICLE AND SINGLE VENTRICLE (?)
PULMONARY STENOSIS

Clinical data: Age 12 years Weight 41 Kg (90¼ lbs) F

History: Cyanosis was first noted when the child began to walk at 1 year Her physical activity has been limited, she fatigues on walking one block

Physical examination: Development was normal Moderate cyanosis was persistent in the nail beds and lips, deepening and becoming widespread with exercise Dyspnea developed after moderate exercise Clubbing was present. The liver edge was felt at the left costal margin

HEART



Size Apex beat just outside right mid clavicular line

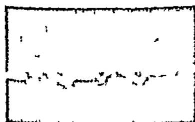
Thrill Systolic maximum in III right interspace

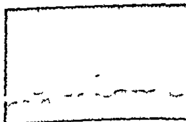
Sounds Second sound sharp and heard equally well on both sides of upper sternum

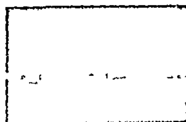
Murmurs 1 Harsh, almost grating systolic, maximum at III right interspace transmitted upward

Arterial blood pressure 92/74 mm Hg.
Hb 19 5 Gm, R B C 7 2 million

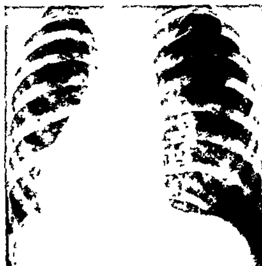
ELECTROCARDIOGRAM


LEAD I


LEAD II


LEAD III

Mirror dextrocardia with "right" preponderance



P A



L A-O



R A-O

The apex was to the right, slightly raised above the diaphragm. The pulmonic area in the postero-anterior view was straight; the pulmonary window in the right anterior-oblique view, however, appeared somewhat clearer than normal Shadows of hilar pulmonary vessels were clearly visible, but no expansile pulsations were present. The lung fields were clear. The ascending aorta was prominent and pulsated very actively. The aortic knob was on the right. In the postero-anterior view a less dense shadow without evident pulsations was seen coursing upward on the left above the ascending aorta. Barium entered the stomach on the right side.

CLINICAL DIAGNOSIS

Dextrocardia with situs inversus Probable tetralogy of Fallot with right aortic arch.

Arterial blood oxygen capacity — 302 cc/lit

Arterial blood oxygen saturation at rest — 79%

Arterial blood oxygen saturation during exercise — 50%

Oxygen intake at rest — 27 1 cc/lit. of ventilation

Oxygen intake during exercise — 20 8 cc/lit. of ventilation

Oxygen consumption at rest = 181 cc/min

ESTIMATED BLOOD FLOW

Systemic (Q_s) — probably equal to or less than 8 60 lit/min

Pulmonary (Q) — 3 70 lit/min

COMMENTS ON THE PHYSIOLOGICAL STUDY

A double superior vena cava was demonstrated by introduction of the catheter down both the left and the right side on two successive occasions (see Figs 17 and 18) The two superior venae cavae communicated with a very large chamber, located posteriorly where the blood pressure was low (see Fig 19) Passing through this chamber the tip of the catheter could be directed from one to the other superior vena cava (see Fig 17), but it could not be seen to enter a pulmonary vein or the inferior vena cava However, it was advanced from the low pressure heart chamber into a cavity where the systolic pressure was very high (see Fig 20), being equal to the systolic pressure in the brachial artery This cavity communicated with the truncus of the pulmonary artery (see Fig 18), in which the systolic pressure was significantly lower Further progress of the tip of the catheter into a right or a left branch of the pulmonary artery was not possible The oxygen content in the first cardiac cavity was only slightly lower than in the second cardiac cavity entered In the latter chamber the oxygen content was identical with that in the pulmonary artery and in the brachial artery Accurate estimation of the systemic flow was not possible in view of (1) the uncertainty concerning the nature of the auricular malformation and (2) the absence of a blood sample from the inferior vena cava the oxygen content of which could have been averaged with that of the superior vena cava The marked reduction both in arterial blood oxygen saturation and in the rate of oxygen removal per liter of ventilation during exercise could be taken as evidence of a greater increase in blood flow through the systemic than through the pulmonary circulation in this state

GENERAL COMMENTS

The data obtained during the physiological studies are consistent with the diagnosis of a single ventricle with pulmonary stenosis The presence of an unsuspected double superior vena cava was clearly demonstrated The existence of a single auricle although not demonstrated directly by passage of the catheter into a pulmonary vein is suspected because the oxygen content in the low blood pressure chamber was higher than in the superior vena cava and only slightly different from that in the single ventricle On the basis of the physiological studies it would seem that the indications for a Blalock Taussig type of operation are not clear cut the pulmonary blood flow was smaller at rest and especially during exercise than the systemic flow however, the blood pressure in the pulmonary artery beyond the stenosis was still elevated

This case of dextrocardia with cyanosis supports the general impression that in such cases multiple and uncommon anomalies are frequently present

BIBLIOGRAPHY

Tl e following list of references l a l een l mited to card ac catl eterization studies in congen tal heart d c e (to Noven l er 1948

Brannon E S Weens H S, and Warren J V Atrial septal defect Study of hemodynamics by *the technique of right heart catheterization An J M Sc 210 480 1945*

Baldwin Eleanor deF Moore L V and Noble R P The demonstration of ventricular septal defect by means of right heart catheterization Am Heart J *32* 152 1946

Dexter L Burwell C S Haynes F W and Seibel R E Venous catheterization for the diag*nosis of congenit il heart di ease Bull New England Med Cent 8 113 1946*

Geiger A J Anderson H C Winkler A W and Kaplan H S Cardiovascular catl eterization as an aid in diagnosis of abnormal cardiova cular communications Connecticut M J *10* 895 1946

Bing R J Vandam L D and Gray F D Jr Physiological tudies in congenital heart disease (I) Procedures Bull Johns Hopkins Hosp *80* 107 1947

Bing R J Vandam L D and Gray F D Jr Physiological studies in congenital heart disease (II) Results of preoperative studies in patients with tetralogy of Fallot. Bull Johns Hopkins Hosp *80* 121, 1947

Sosman M C Venous catheterization of the l eart. (I) Indications technics and errors Radiology *48* 441 1947

Dexter L Venous catheterization of the heart (II) Results interpretations and value Radiology *48* 451 1947

Dexter L Haynes F W Burwell C S Eppinger E C Seibel R E and Evans J M Studies of congenital heart disease (I) *Technique of* venous catheterization as a diagnostic proce dure J Clin Inves *26* 547 1947

Dexter L. Haynes F W Burwell C S Eppinger E C Sagerson R P and Evan J M Studies of congenital heart disease (II) The pressure and oxygen content of blood in the right auricle right ventricle and pulmonary artery in control patients with ob ervations on the oxygen saturation and source of pulmonary capillary blood J Clin Inve *26* 554 1947

Dexter L Haynes F W Burwell C S Eppinger E C Sosman M C and Evans J M Studies of congenital heart disease (III) Venous catheterization as a diagno tic aid in patent ductus arteriosus tetralogy of Fallot ventricular septal defect and auricular septal defect J Clin Inves *26 561 1947*

Johnson *A L Wollin D G and Ro* J B Heart catheterization in the inve tigation of congenital heart disease Canad M A J *56* 249 1947

Bing R J Vandam L. D and Gray F D Jr Physiological studies in congenital heart disease (III) Results obtained in five ca es of Eisenmenger s complex. Bull Johns Hopkins Hosp *80* 323 1947

Cournand A Cardiac catheterization in the diagno is of congenital heart disease a report. International Congre s of Pediatric N Y 1947

Vandam, L D, Bing R J, and Gray, F D, Jr, Physiological studies in congenital heart disease (iv) Measurements of the circulation in five selected cases Bull Johns Hopkins Hosp *81* 192, 1947

Cournand, A, Motley, H L, Himmelstein, A, Dresdale, D, and Baldwin, J, Recording of blood pressure from the left auricle and the pulmonary veins in human subjects with inter auricular septal defect Am J Physiol *150* 267, 1947

Massee, J C Atrial septal defect correlation of necropsy with data obtained by right heart catheterization Am J Med Sc *211* 248, 1947

Howarth, S, McMichael J, and Sharpey Schafer, E P, Cardiac catheterization in cases of patent interauricular septum, primary pulmonary hypertension, Fallot tetralogy, and pulmonary stenosis Brit Heart J *9* 292, 1947

Burchell H B Parker, R L Dry, T J Wood, E H, Pender, J W, and Pugh, D G, Cardiac catheterization in the diagnosis of various cardiac malformations and diseases Proc. Staff Meet Mayo Clin *23* 481, 1948

Taylor B E Geraci, J E, Pollack, A A, Burchell H B, and Wood, E H, Interatrial mixing of blood and pulmonary circulatory dynamics in atrial septal defects Proc Staff Meet, Mayo Clin *23* 500 1948

DuShane J W and Montgomery G E, Jr, Patent ductus arteriosus with pulmonary hypertension and atypical clinical findings Proc Staff Meet, Mayo Clin *23* 505, 1948

Burchell H B Taylor B E Pollack A A, DuShane J W, and Wood, E H, Ventricular septal defect and pulmonary hypertension without hypoxemia Proc Staff Meet, Mayo Clin *23* 507, 1948

Geraci J E Dry T J, and Burchell, H B, Atrial septal defect and probable tricuspid atresia in adults Proc Staff Meet Mayo Clin *23* 510 1948

Pollack A A Taylor, B E Odel H M and Burchell H B, Pulmonary stenosis without septal defect Proc Staff Meet Mayo Clin *23* 516, 1948

Keith J D, Diagnosis of congenital heart disease Canad M A J *58* 247, 1948

Handelsman, J C Bing R J Campbell, J A, and Griswold, H E, Physiological studies in congenital heart disease (v) The circulation in patients with isolated septal defects Bull Johns Hopkins Hosp *82* 615, 1948

Combined Staff Clinics—College of Physicians and Surgeons, Columbia University and Presbyterian Hospital New York, Congenital heart disease Am J Med *4* 886 1948

Greene, D G Baldwin Eleanor deF, Baldwin, Janet S Himmelstein, A, Roh, C E, and Cournand A, A report of four cases of pure congenital pulmonary stenosis and of four cases of idiopathic congenital dilatation of the pulmonary artery, with a review of the literature Am J Med (in press)